

BAV et stimulation

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www.Rhythmopedia.com

ECG NORMAL
ET PATHOLOGIQUE

CARDIOPATHIES
CONGÉNITALES

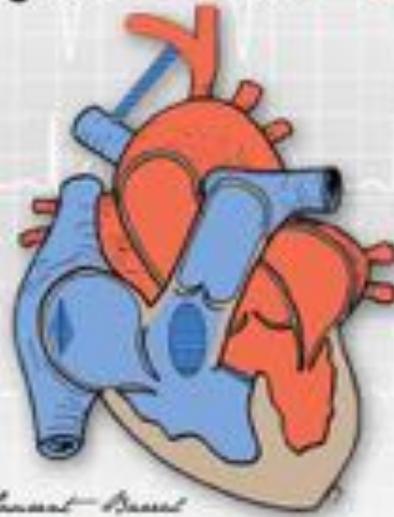
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avec le groupe de
Réflexion de
Rythmologie
Pédiatrique
et Congénitale



Formation complète
à l'ECG de l'enfant
et de l'adulte jeune

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RECHERCHER UN TRACÉ

Titre

Cardiopathies

-Tout-



Rechercher

<http://www.rhythmopedia.com>

Etiologies

Brady-arythmie de l'enfant

**Cœur
anatomiquement
sain**

BAV Immun

BAV progressif

Canalopathie

Maladie rythmique de
l'oreillette

Dysfonction sinusale

QT Long

**Cardiopathie
congénitale**

Pré-opératoire

BAV « malformatif »

Dysfonction sinusale/hétérotaxie

Post-opératoire

BAV post-opératoire

Maladie rythmique de l'oreillette

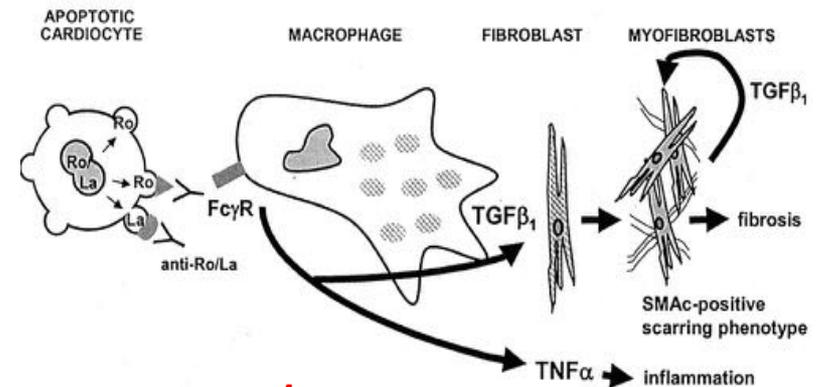
Dysfonction sinale/Senning-Mustard

BAV Complet

BAVc de diagnostic périnatal

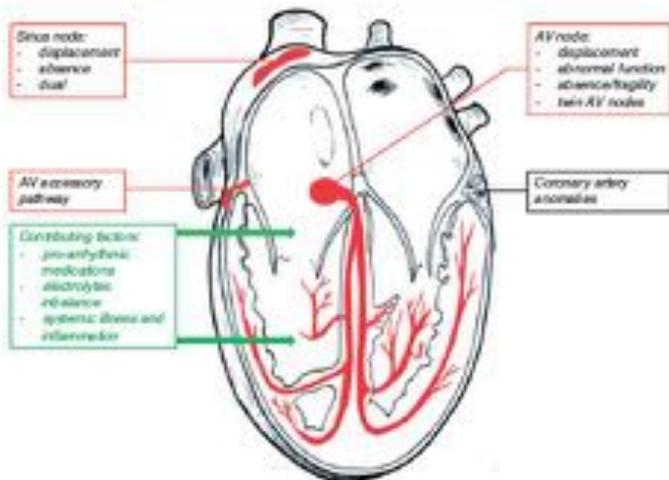
- BAV « malformatif »
 - Anomalie de la jonction auriculo-ventriculaire
 - 40% de BAVc après 20 ans

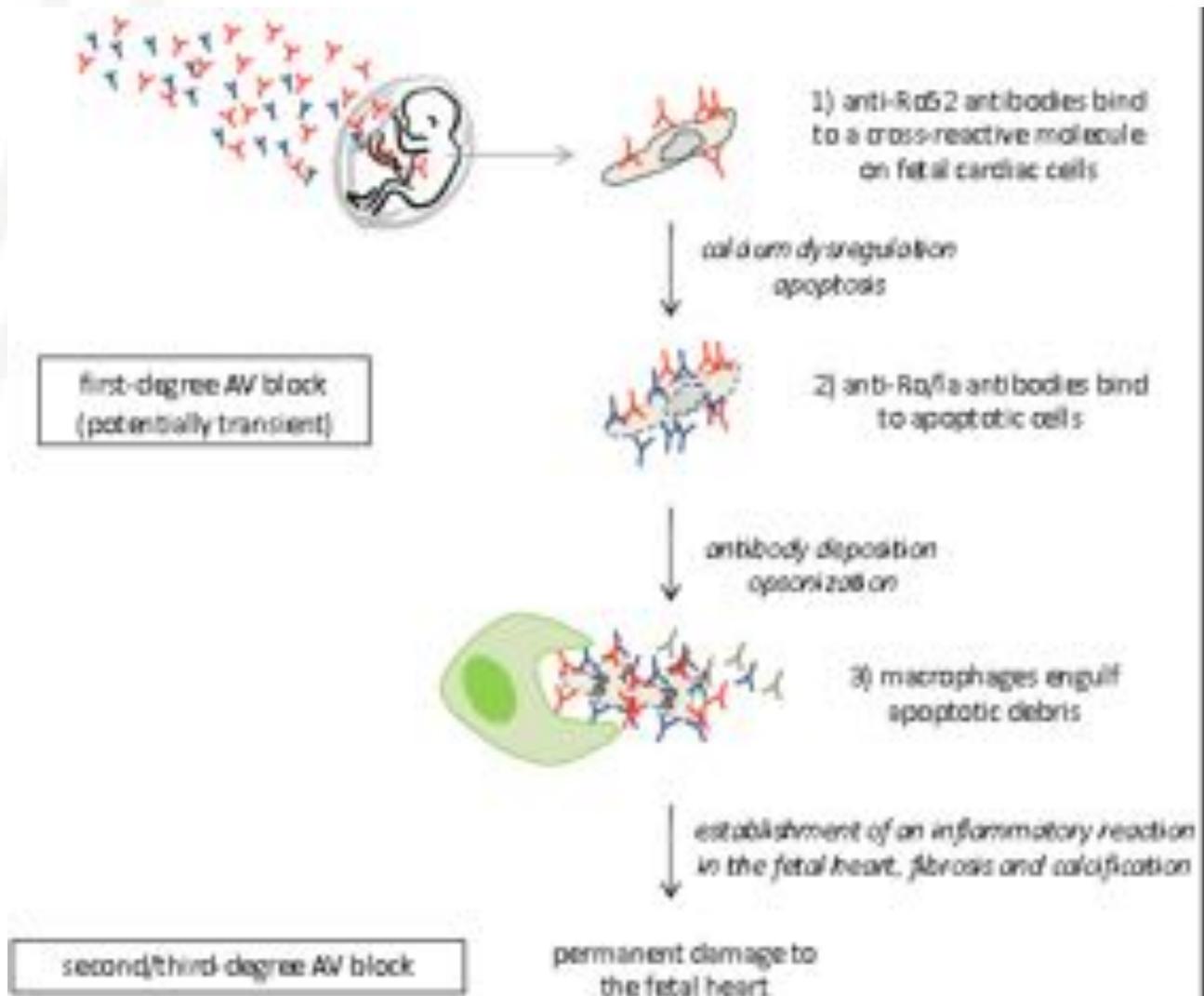
- BAV « immun »



- **AC anti SSA/SSB**

- Diagnostic entre 16 et 24 SA
- 1/10 000 NN vivants
- Mortalité périnatale: 16-19%
- Risque de 2 à 3% pour Primipare
- Risque de récurrence de 20% =>Protocole PLAQUENIL®
- **Risque de cardiomyopathie II^{aire} +++**

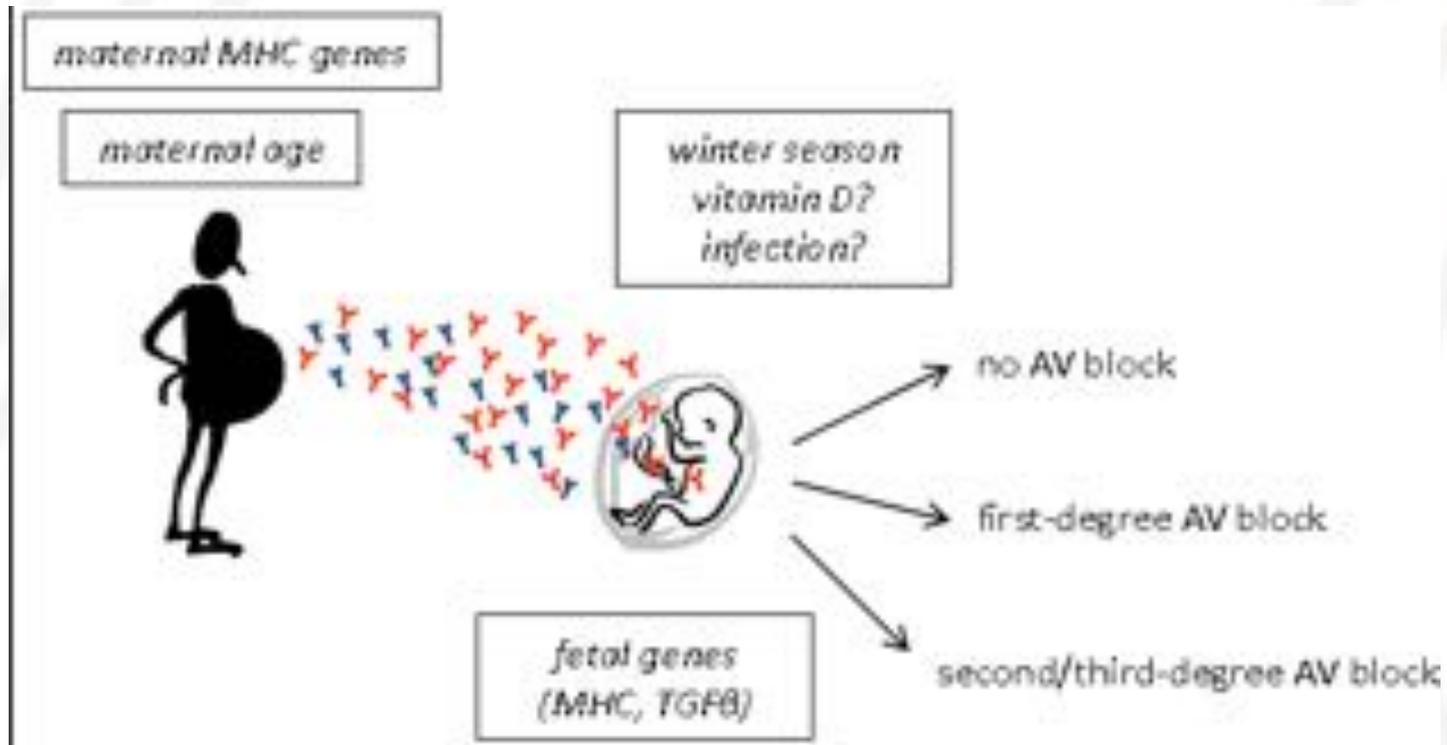




- Modele affection auto immune acquise “passiive”
- (1st, 2nd, 3rd-degree heart block) most commonly develop during **18–24 weeks** of pregnancy.
- CHB is a **progressively developing disease** and 3rd-degree heart block appears to be irreversible

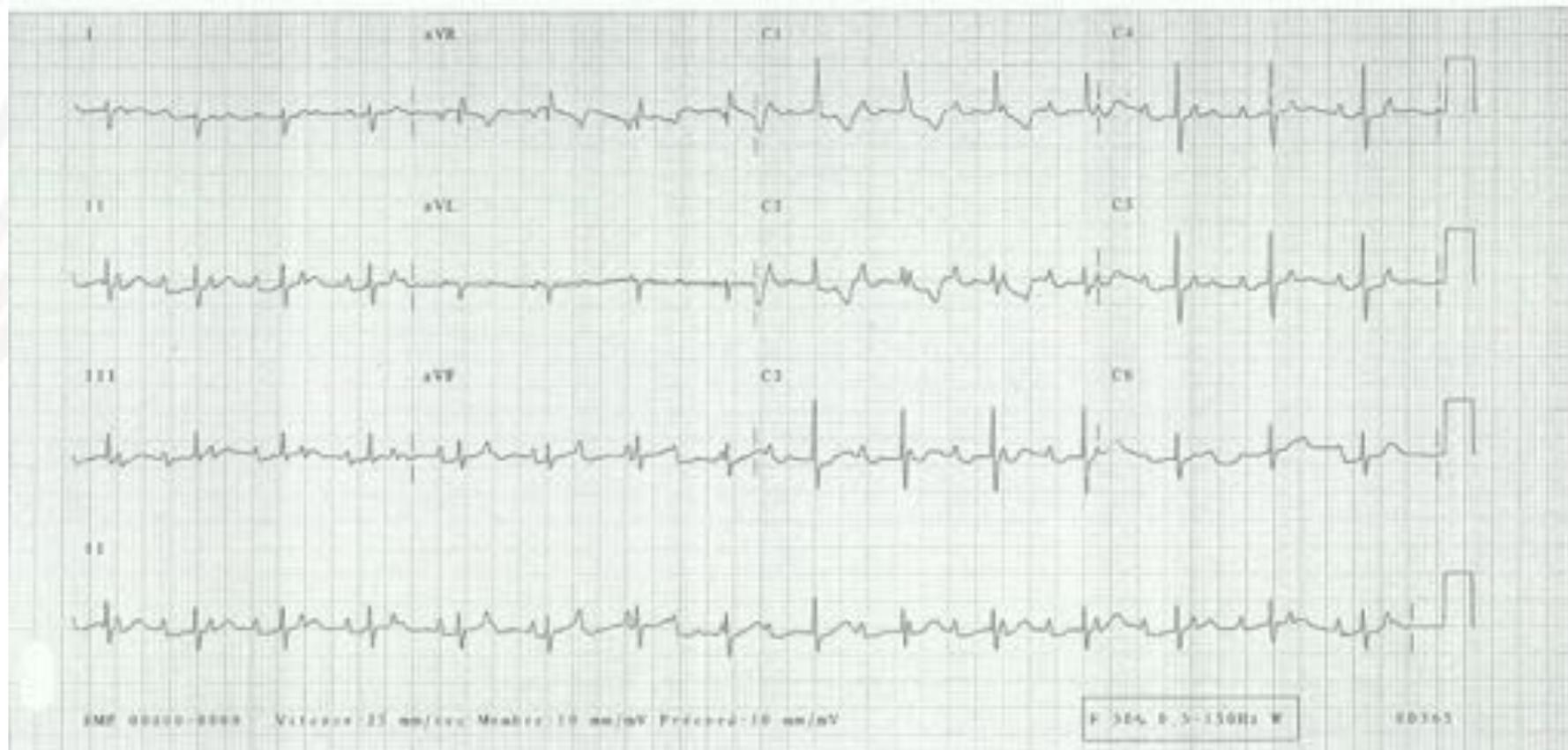
Background

- 85% of foetus with congenital heart block and absence of structural abnormalities have maternal transfer of antibodies against SSA/Ro and SSB/La
- however only 2% of seropositive mother have newborns with congenital heart block
- This low risk rate rises to 19% for women with a previously affected newborn



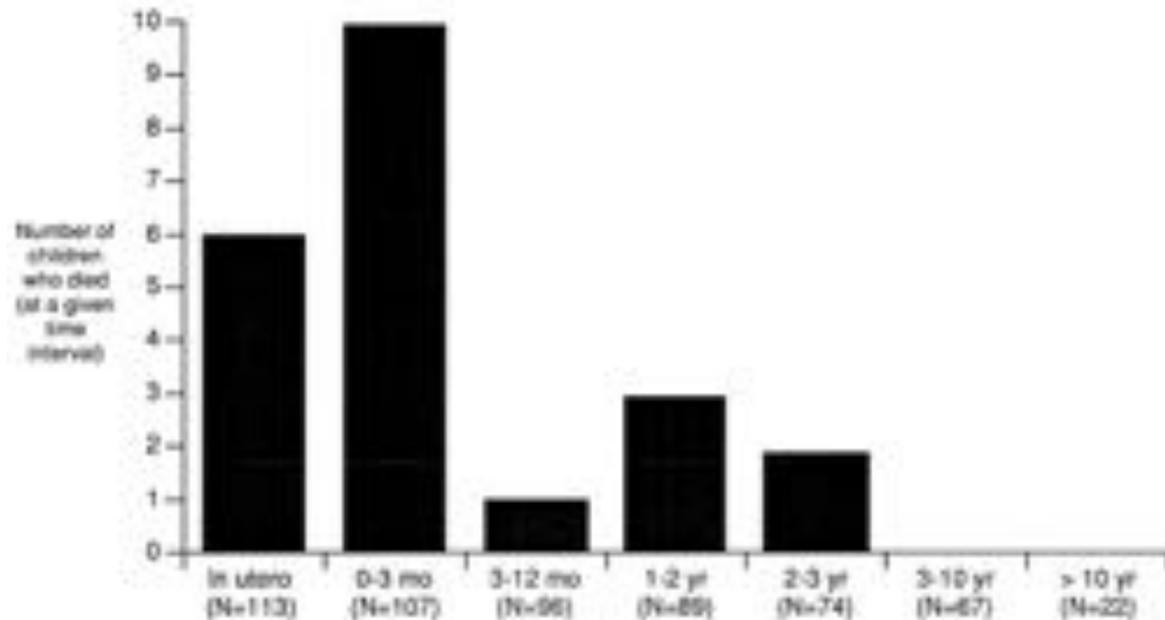
- antibodies to SSA/Ro and SSB/La could not be **the only cause of the disease** and other maternal and foetal factors are important.
- Nevertheless, maternal health status is not considered a risk factor for CHB; approximately **40-60% of mothers with an affected newborn are totally asymptomatic** for autoimmune disease when foetal bradycardia is found

BAVc lupique néonatal



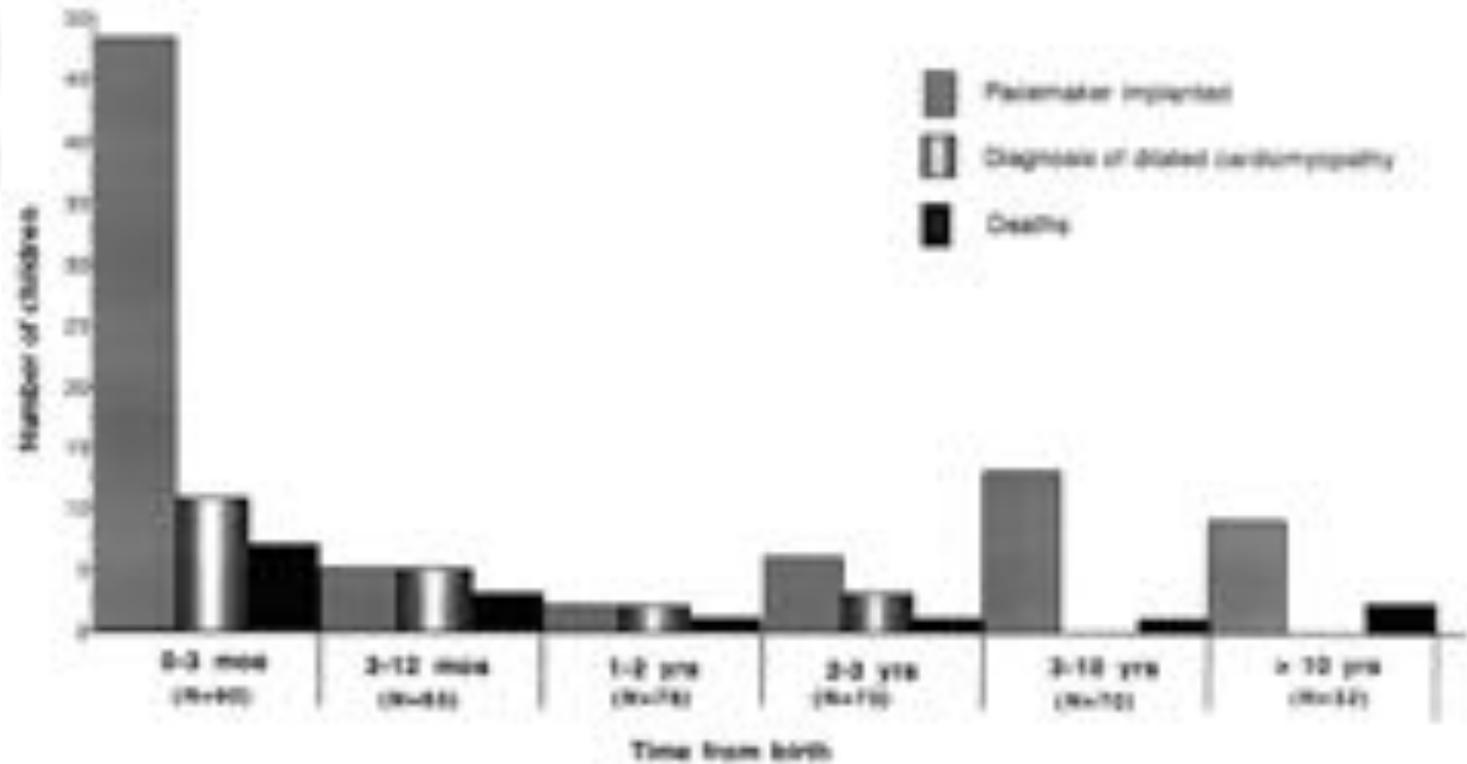
Pronostic

- despite early diagnosis and treatment CHB carries high fetal and neonatal mortality
- mortality rate varied from 8 % to 19 % despite early pacing
- mostly attributable to cardiac failure in the neonatal period
- Importance of the inflammatory process
- Fetal hydrops, fibroelastosis, < 32 weeks = Poor prognosis

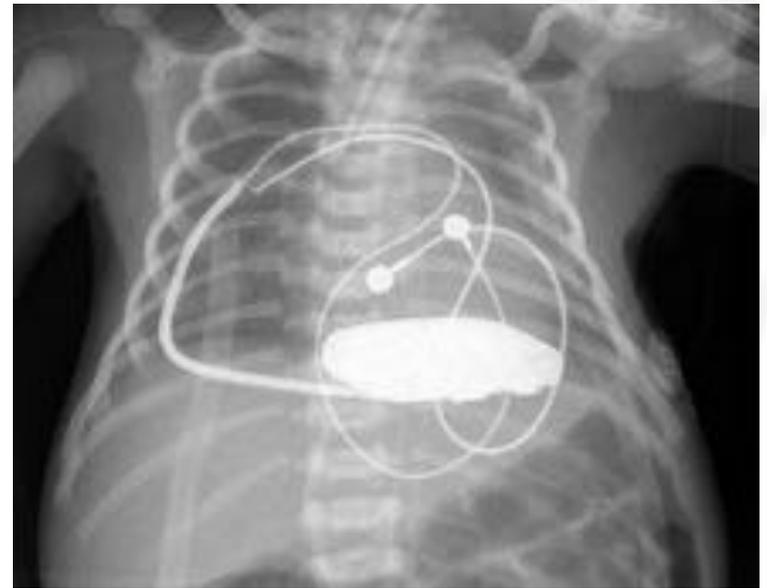


Buyon J et al; JACC 1998

Prognosis



Treatment



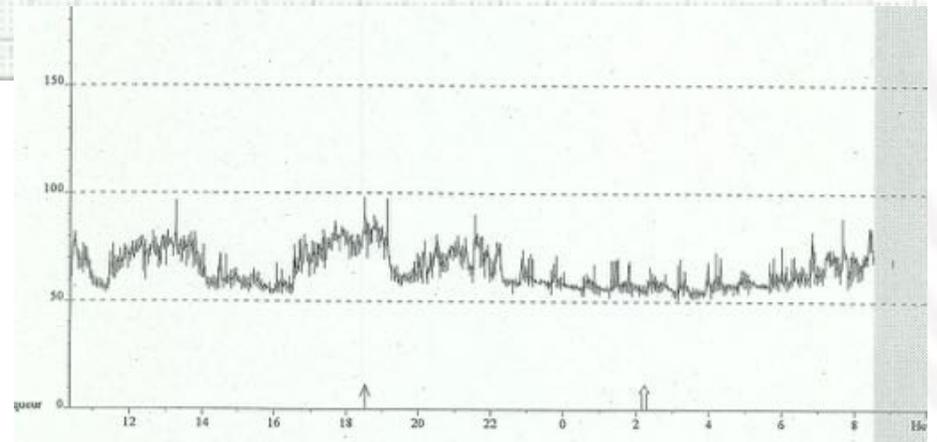
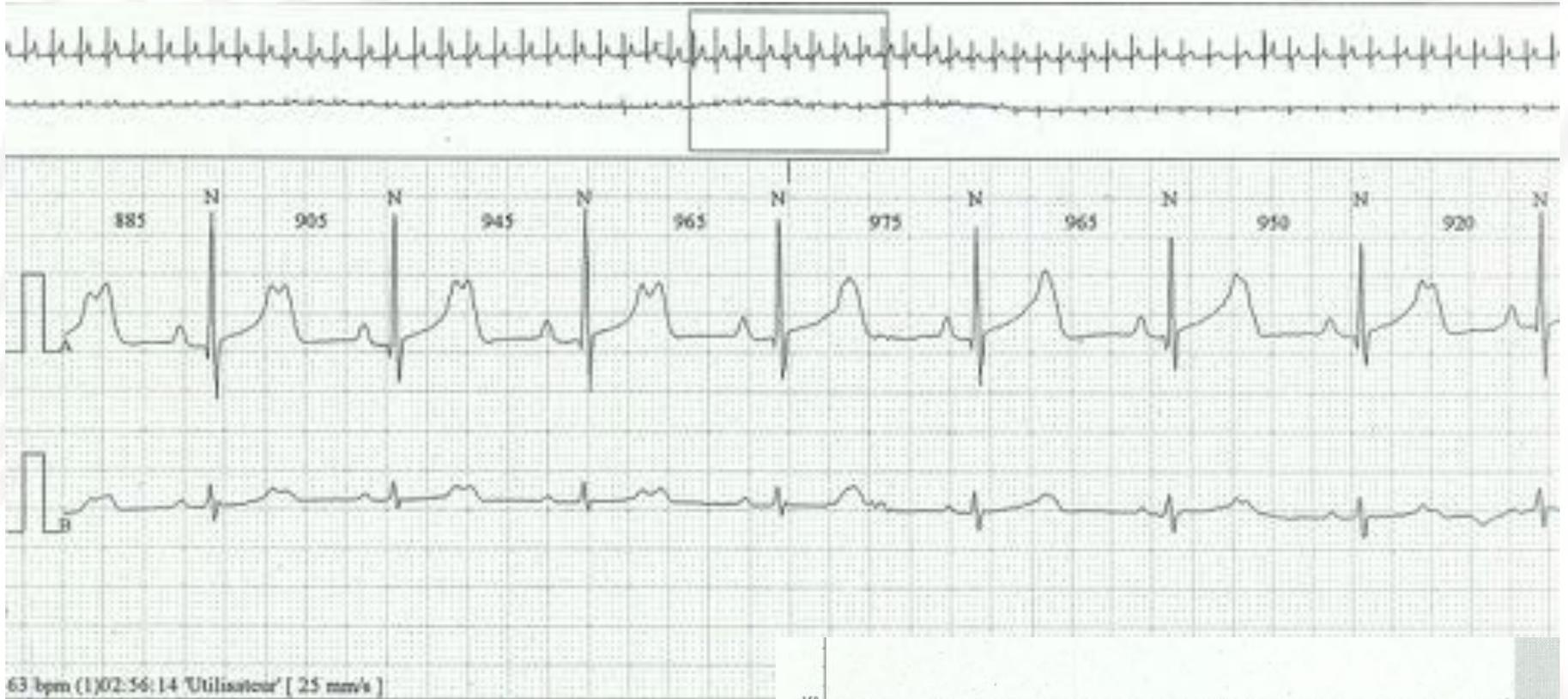
- Risk for syncope, heart failure and sudden death is present at any age including fetal life.
- Only treatment is **pacemaker implantation (PMI)**
- Specific pacing indication for asymptomatic patients



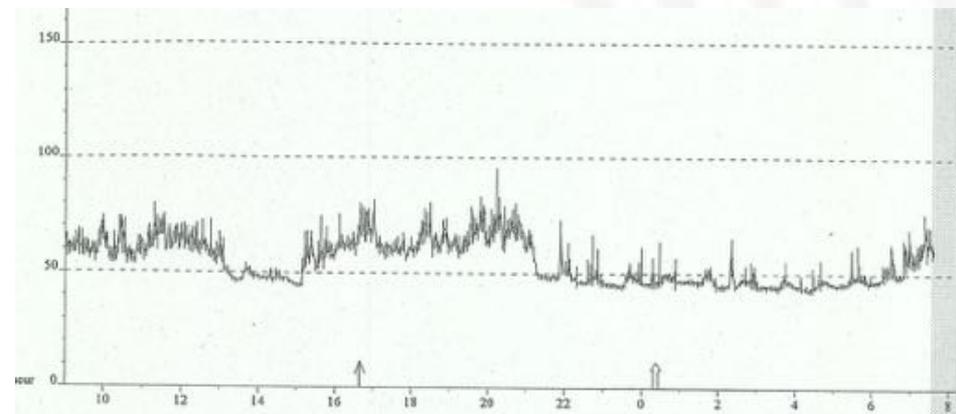
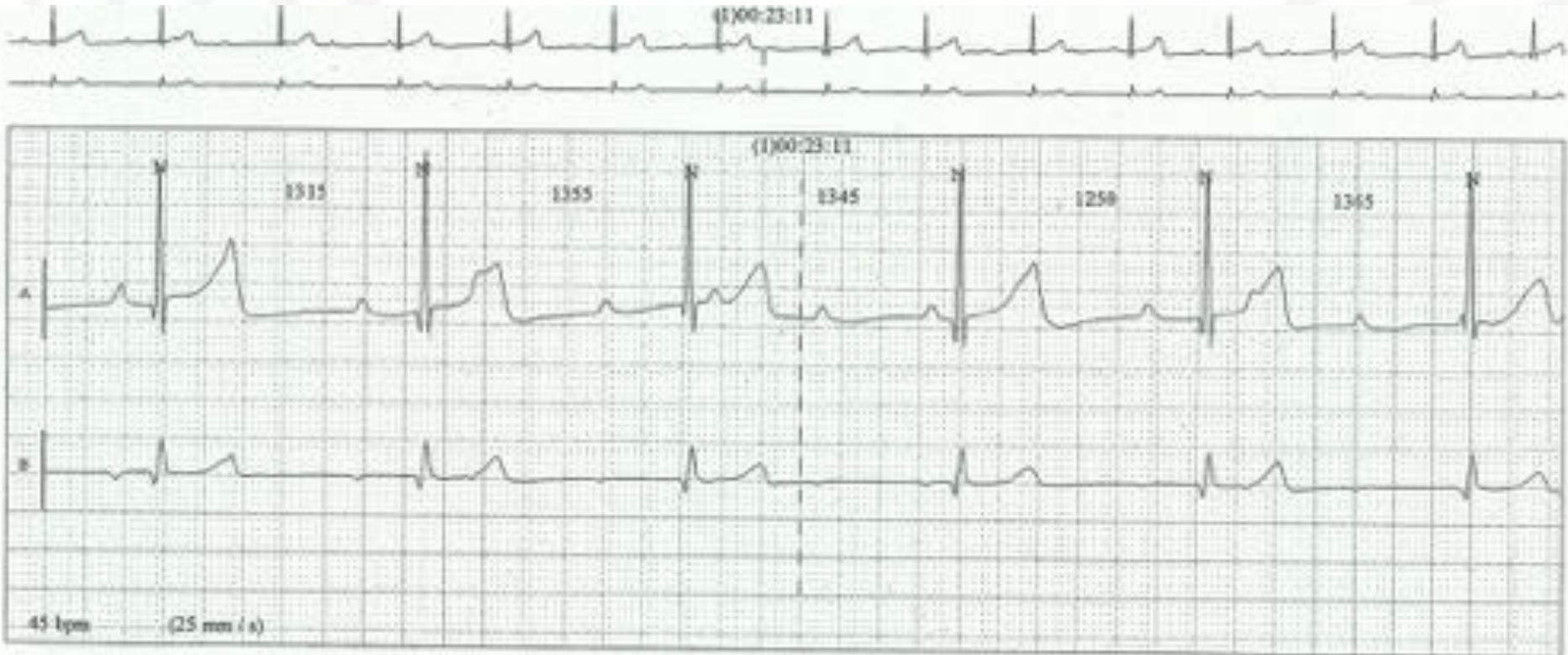
BAVc de l'enfance

- « Progressive conduction cardiac disease » (PCCD)
 - Cause inconnue le plus svt
 - Absence anticorps maternels
 - Souvent après 6 mois
 - SCN5A, SCN1B (Brugada-Ac/Aa...)/GJA5
 - Forme familiale: Chromosome 19, TRPM4, NKX2.5 (+ CIA ostium secundum)
- Blocs infectieux et toxiques
- Associé à des myopathies et cardiomyopathies
 - Steinert
 - Sd de Kearns-Sayre
 - Laminopathie, desminopathie

BAVc enfance



Décembre 2010



Mai 2011

INDICATIONS DE STIMULATION

BAV complet

Tableau 1 Recommandations issues des sociétés européennes et américaines de cardiologie sur l'implantation de pacemakers en pédiatrie.

Indication	Classe	Niveau de preuve
Une stimulation cardiaque permanente est indiquée en cas de BAV complet associé avec l'un des éléments suivants : symptômes, dysfonction ventriculaire, intervalle QTc prolongé, extrasystoles ventriculaires complexes, rythme d'échappement à QRS larges, fréquence cardiaque < 55 bpm chez le nourrisson, fréquence cardiaque < 70 bpm dans le cadre d'une cardiopathie congénitale, pauses cardiaques > 3 fois le cycle de base.	I	B
Une stimulation cardiaque permanente est indiquée en cas BAV du 2 ^e ou du 3 ^e degré associé à des symptômes ou à une dysfonction ventriculaire.	I	C
Une stimulation cardiaque permanente est indiquée en cas BAV du 2 ^e ou du 3 ^e degré survenant en postopératoire et persistant plus de 7 jours après la chirurgie.	I	C
Une stimulation cardiaque permanente est indiquée en cas de syndrome du QT long associé à l'un des éléments suivants : BAV en 2/1 ou du 3 ^e degré, bradycardie symptomatique (spontanée ou due aux bêtabloquants), tachycardie ventriculaire pause dépendante.	IIa	B
Une stimulation cardiaque permanente est indiquée en cas cardiopathie congénitale avec altération de l'hémodynamique due à une bradycardie sinusale ou à un asynchronisme atrioventriculaire.	IIa	C
Une stimulation cardiaque permanente est à considérer en cas de BAV congénital du 2 ^e ou du 3 ^e degré en l'absence de symptômes, dysfonction ventriculaire, intervalle QTc prolongé, extrasystoles ventriculaires complexes, rythme d'échappement à QRS larges, fréquence cardiaque < 55 bpm chez le nourrisson, pauses cardiaques > 3 fois le cycle de base.	IIb	B
Une stimulation cardiaque permanente est à considérer en cas de pathologie neuromusculaire associée à un bloc auriculo-ventriculaire asymptomatique quel qu'en soit le degré.	IIb	C
Une stimulation cardiaque n'est pas recommandée en cas de BAV du 1 ^{er} degré ou du 2 ^e degré de type Luciani-Wenckebach asymptomatiques.	III	C

2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy

The Task Force on cardiac pacing and resynchronization therapy of the European Society of Cardiology (ESC). Developed in collaboration with the European Heart Rhythm Association (EHRA).

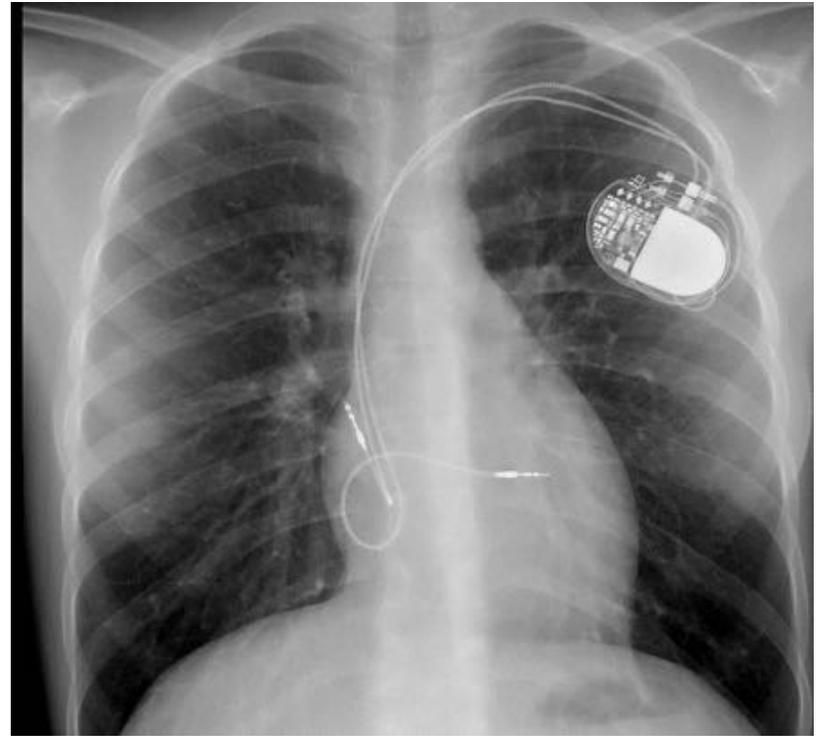
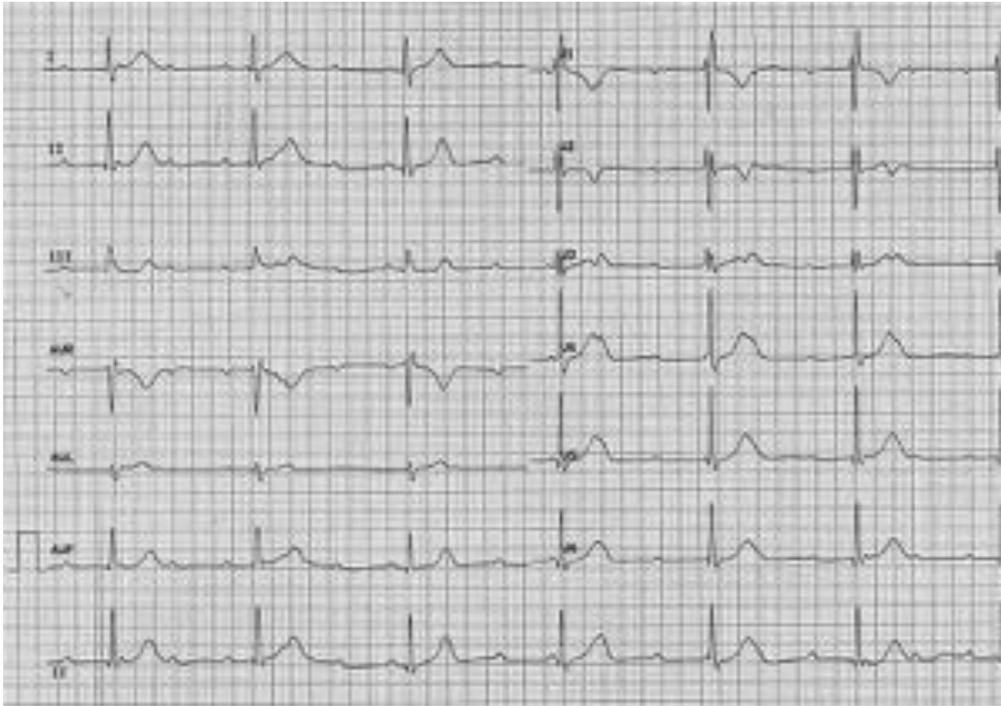
1. Congenital third-degree atrioventricular block with any of the following conditions: Symptoms Ventricular rate <50–55/min in infants Ventricular rate <70/min in congenital heart disease Ventricular dysfunction Wide QRS escape rhythm Complex ventricular ectopy Abrupt ventricular pauses >2–3 × basic cycle length Prolonged QTc Presence of maternal antibodies-mediated block ^{188–198}	Class I	B
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2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy

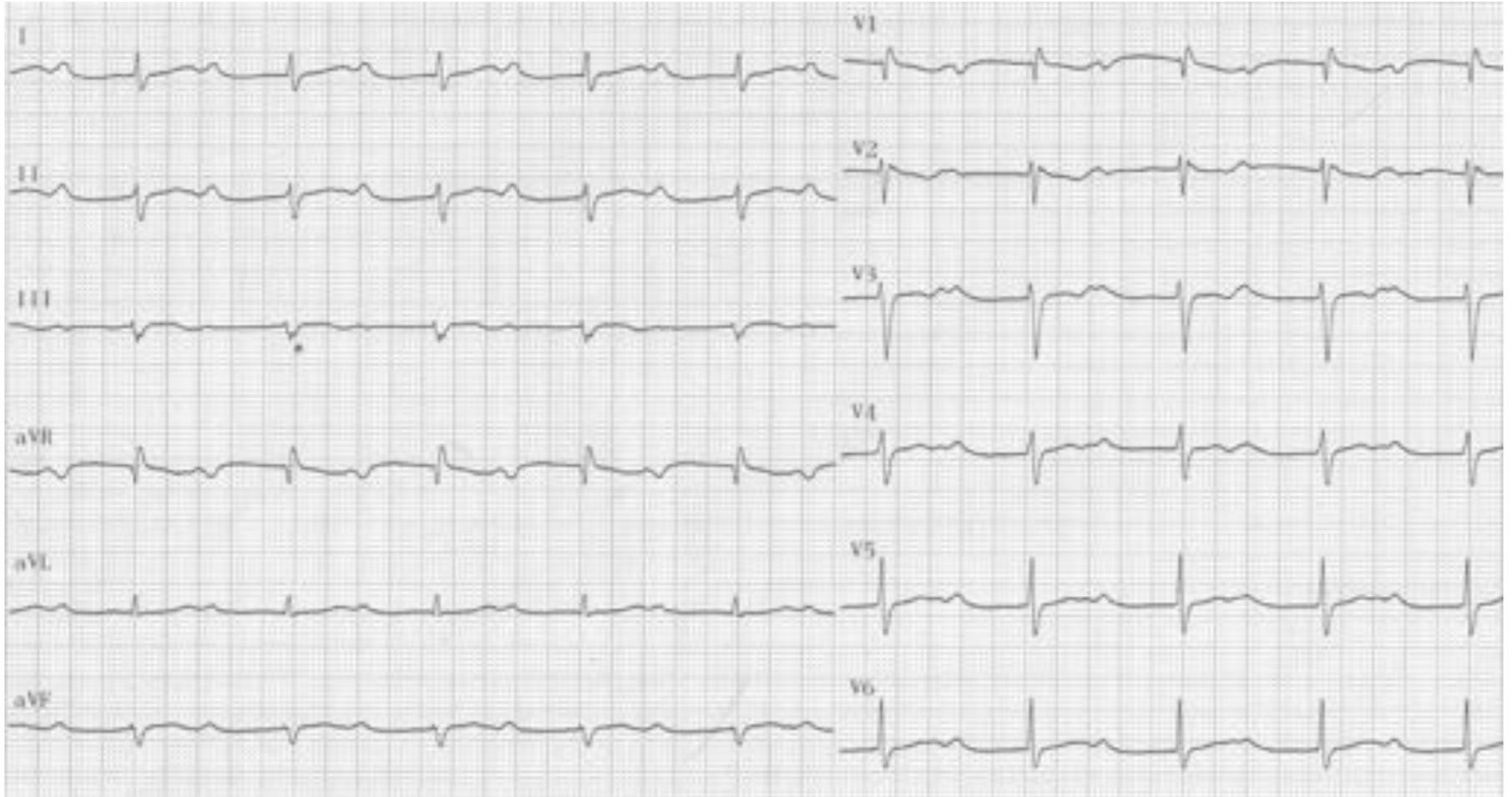
The Task Force on cardiac pacing and resynchronization therapy of the European Society of Cardiology (ESC). Developed in collaboration with the European Heart Rhythm Association (EHRA).

Congenital third-degree atrioventricular block without a Class I indication for pacing ^{188–198}	Class IIb	B
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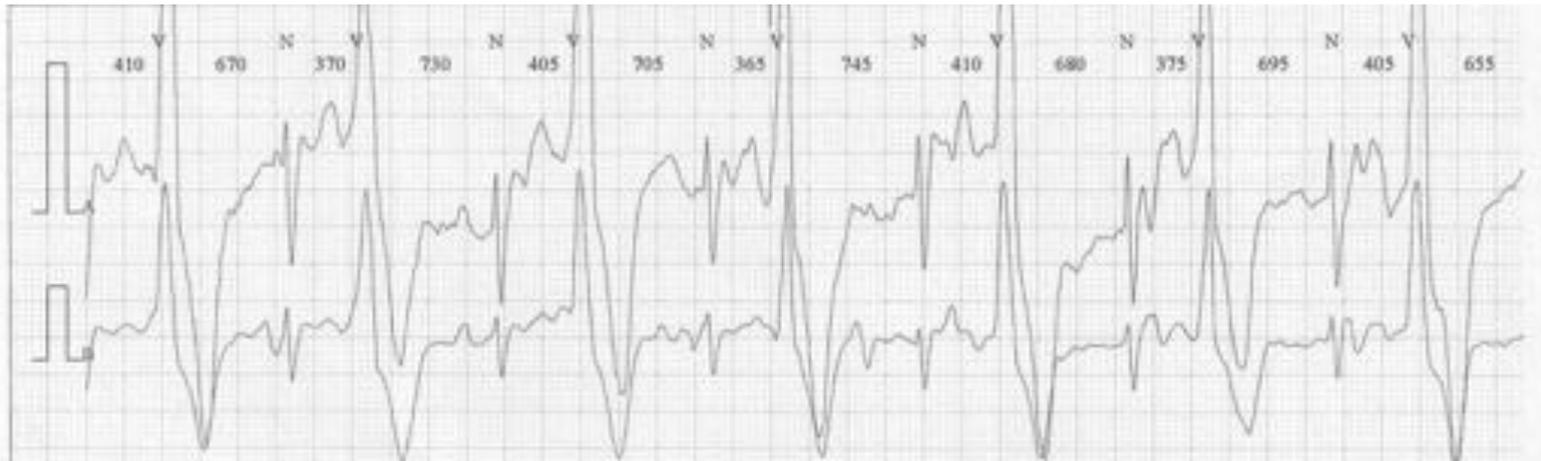
Patiente 16 ans: syncope



Patiente 25 ans: asymptomatique



Patiente 25 ans: asymptomatique



BAV complet: facteurs de risque

Karpawich 1980	24 pts fu : 5 yrs	7 syncope	1 SD	FC < 50
Reid (1982)	35 pts (21 PM)	1 syncope	1 SD	FC < 50
Dewey (1987)	27 pts (holter)	0 3 syncope	0 2 SD	FC > 50 FC < 50
Batisse	39 pts	6 syncope	2 SD	FC < 50 < 2 ans < 45 (2-4 ans) < 40 (> 4 ans)
Michaelsson 1995	302 pts			FC < 55 bpm

Indications for permanent pacing in children

Absolute indication

Syncope attacks
Congestive heart failure
L Ventricular dysfunction

Gregoratos G, Circulation 2002

High risk

< 2 years :

HR < 55 beats/min in isolated block
HR < 70 beats/min with associated disease

> 2 years :

Mean HR < 50 beats/min
HR < 40-45 beats/min
Abrupt pauses in ventricular rate (3 times basic cycle length)

All patients older than 15 years

Michaelson, Pace 1997

Wide QRS escape rhythm (QRS>120ms) or ventricular dysfunction, prolonged QT interval, ESV

BAVc/indications de stimulation

- BAVc symptomatique

- Défaillance cardiaque
- Syncope
- Intolérance à l'effort
- Mauvaise prise de poids

- Dysfonction VG/ETT

- DTDVG ↗ mais FEVG #

- Implantation prophylactique

- Rythme d'échappement lent
 - Avant 2 ans : **≤ 50-55 bpm** ,
< 70 si cardiopathie associée
 - Après 2 ans : **FC moyenne < 50 bpm** ,
Fc instantanée < 45
ou pause > 2-3 X RR de base
- Anomalie ECG/Holter
 - QRS d'échappement larges
 - ESV, TDR ventriculaire
 - QT allongé

Troubles conductifs post opératoires

Case	Heart disease	Procedure and timing (age [y] and year of surgery)	Chronic postsurgical AV block	Time to recovery of conduction	Follow-up*
1	AVSD	AVSD correction (4.0 y; 2006)	Complete	22 d; 4.7 y	Asymptomatic first-degree AV block; completely normal AV conduction 4.6 y later (5.1 y)
2	D-TGA, VSD, PS	VSD closure (2.6 y; 2007)	Complete	23 d	Normal AV conduction until death (<i>Klebsiella sepsis</i>) (2 mo)
3	Isolated VSD	VSD closure (5.9 y; 2003)	Complete	11 wk	Normal AV conduction during 3.5 y; subsequent relapse into complete AV block (7 y)
4	AVSD	AVSD correction (6.2 y; 1996)	Second degree (advanced)	3.1 y	Asymptomatic first-degree AV block (3.5 y)
5	AVSD	AVSD correction (0.6 y; 2003)	Second degree (advanced)	6.2 y; 7.3 y	Asymptomatic first-degree AV block; completely normal AV conduction 1.1 y later (7.8 y)
6	AS (bicuspid)	LVOT (Ross) procedure (4.5 y; 2004)	Complete	6.8 y	Normal AV conduction (11 mo)
7	AVSD	AVSD correction (0.3 y; 2000)	Complete	7.0 y	Normal AV conduction (3.9 y)

- Double switch (15,6%), Tricuspid (7,8%) et Mitrale (7,4%) remplacement, Switch Atrial avec CIV (6,4%), Rastelli (4,8 %),
- Fermeture CIV par voie percutanée (3-20%)
- Survient immédiatement après chirurgie ou tôt dans période post-op
- Rares cas de survenus après des mois ou années.

Mortalité des BAV post-opératoires persistants NON appareillés

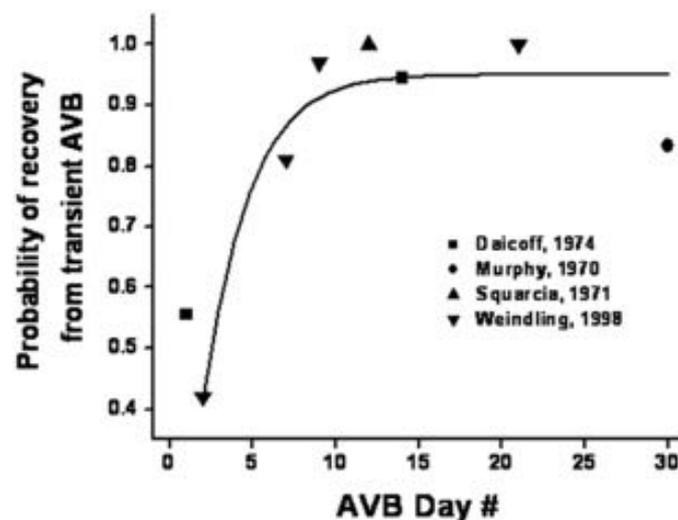
Taux de Mortalité	Période	Référence
100%	Avant 1956	Lillehei et al.
80%	1956-1961	Lillehei et al.
30%	1956-1966	Hurwitz et al.
38%	1957-1973	Hofschire et al.
28%	1957-1978	Driscoll et al.
100%	1960-1967	Squarcia et al.
64%	1962-1968	Murphy et al.

BAV transitoire / BAV persistant

- 2/3 des BAV post op sont transitoires
- Délai de « guérison »
 - 95% avant 9 jours
 - 9% de trouble conducteur partiel résiduel

Weindling et al. Am J Cardiol, 1998

Gross et al. Heart Rhythm, 2006



- Récupération tardive
 - 32% des patients implantés retrouvent une conduction AV complète ou partielle à 5.5 ans (0.1-20)
- (ré)apparition tardive de BAV
 - de 2 mois à 19 ans
 - 23% de BAV transitoire péri-op

Bruckheimer et al.

J Interv Card Electrophysiol, 2002

Lieberman et al. Pediatr Cardiol, 2008

Late recovery of atrioventricular conduction after postsurgical chronic atrioventricular block is not exceptional

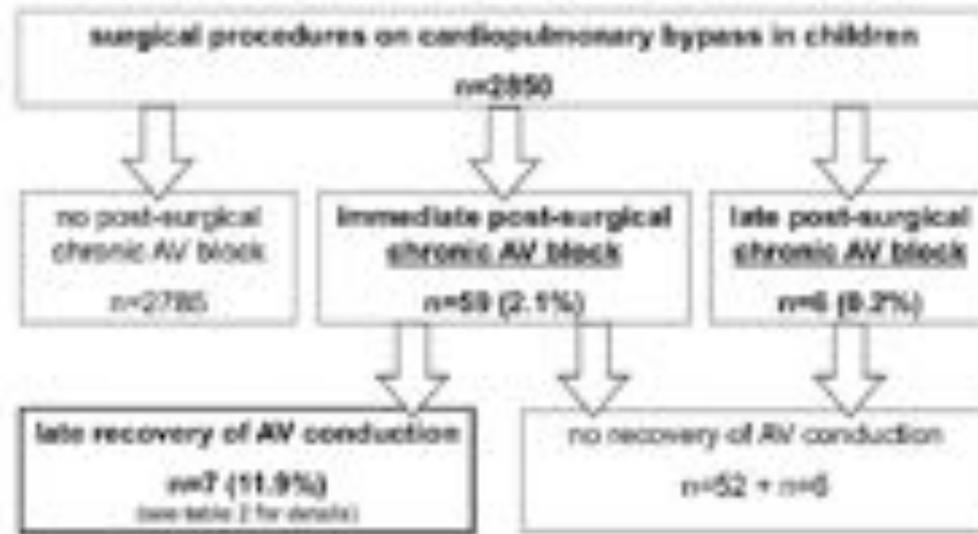


FIGURE 1. Schematic overview of results. Chronic atrioventricular (AV) block was considered to be atrioventricular block of advanced second or third degree persisting longer than 14 days.

2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy

The Task Force on cardiac pacing and resynchronization therapy of the European Society of Cardiology (ESC). Developed in collaboration with the European Heart Rhythm Association (EHRA).

Authors/Task Force Members: Michele Brignole (Chairperson) (Italy)*, Angelo Auricchio (Switzerland), Gonzalo Baron-Esquivias (Spain), Pierre Bordachar (France), Giuseppe Boriani (Italy), Ole-A Breithardt (Germany), John Cleland (UK), Jean-Claude Deharo (France), Victoria Delgado (Netherlands), Perry M. Elliott (UK), Bulent Gorenek (Turkey), Carsten W. Israel (Germany), Christophe Leclercq (France), Cecilia Linde (Sweden), Lluís Mont (Spain), Luigi Padeletti (Italy), Richard Sutton (UK), Panos E. Vardas (Greece)

3) Postoperative AV block in congenital heart disease. Permanent pacing is indicated for postoperative advanced second degree or complete AV block persisting >10 days.	I	B	137-141
4) Postoperative AV block in congenital heart disease. Permanent pacing should be considered for persistent, asymptomatic post-surgical bifascicular block (with or without PR prolongation) associated with transient, complete AV block.	IIa	C	-

EN PRATIQUE, au-delà de J10/J15...

- BAV de haut degré, BAV complet
 - PM sans explo **Class I**
- BAV < 48 heures
 - Pas de PM, pas d'explo **Class III**
- 48 h < BAV < 10 jours, ECG strictement superposable à l'ECG pré-op
 - Pas de PM, pas d'explo **Class III**
- 48 h < BAV < 10 jours mais persistance de trouble conducteur partiel et/ou bloc fasciculaire
 - **Explo EE avec mesure du HV**

D'après Villain et al. Arch Mal Cœur, 2003

Pediatric Post-Operative Atrio-Ventricular Block Meets the Affordable Care Act: A New Strategy for Management

Open Journal of Pediatrics, 2017, 7, 118-127

<http://www.scirp.org/journal/ojped>

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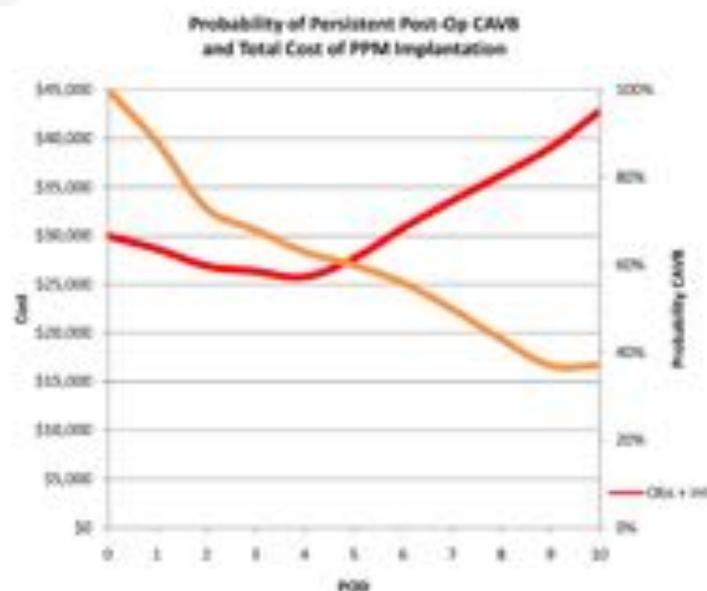


Figure 1. Cost comparison between patients with post-op CHB who are either observed for recovery of native conduction (orange line) or who have a PPM implanted on the corresponding POD due to persistence of CAVB (red line), from POD 0 - 10. CAVB = complete atrioventricular block; PPM = permanent pacemaker; POD = post-operative day.

Table 1. Cost of PPM placement for persistent CAVB from POD 0 - 10.

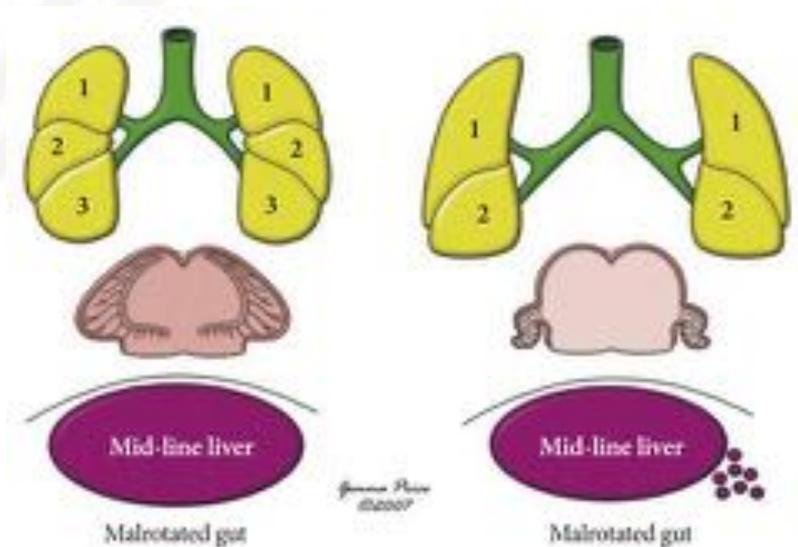
POD	Probability post-op CAVB	Minimum Total Cost
0	1	\$29,342
1	0.88	\$27,823
2	0.73	\$25,623
3	0.68	\$25,237
4	0.63	\$24,899
5	0.6	\$24,628
6	0.56	\$24,903
7	0.5	\$22,737
8	0.43	\$21,201
9	0.37	\$19,416
10	0.37	\$40,312

Model assumes that only patients with the probability of persistent CAVB receive PPMs. CAVB = complete atrioventricular block; PPM = permanent pacemaker.

Bradycardie sinusale et Maladie rythmique de l'oreillette

Dysfonction sinusale « native »

- Hétérotaxie, Lévo-isomérisme



retours veineux

VCS gauche



anatomie des ventricules

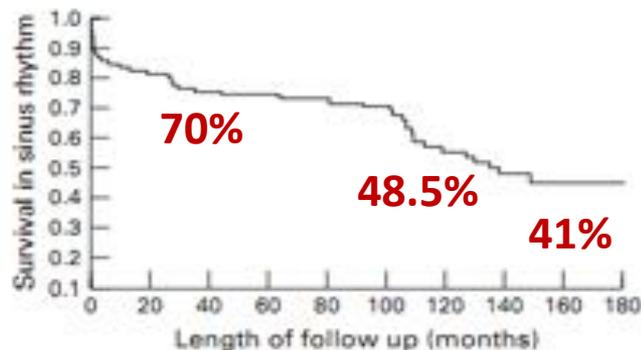
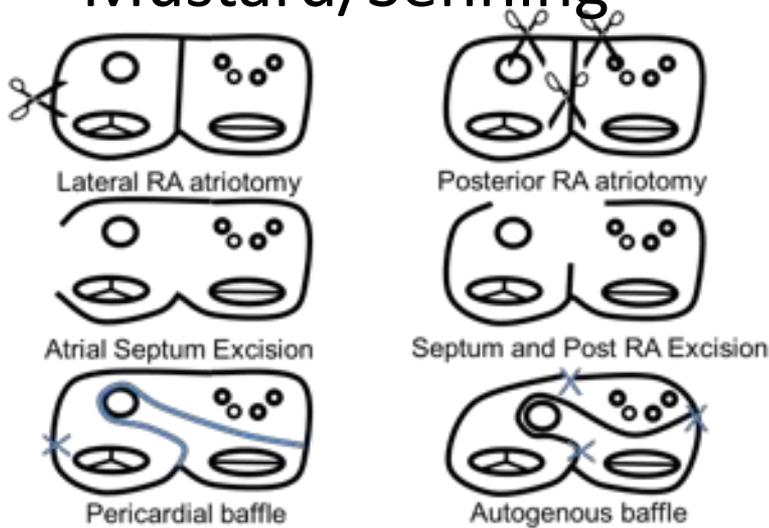
CIV

VU

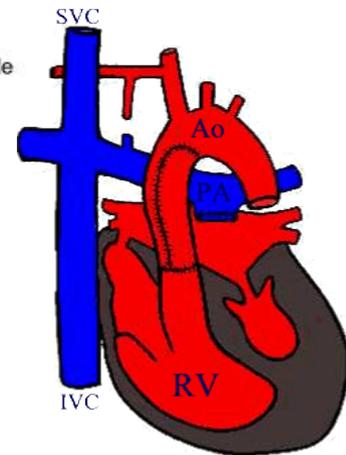
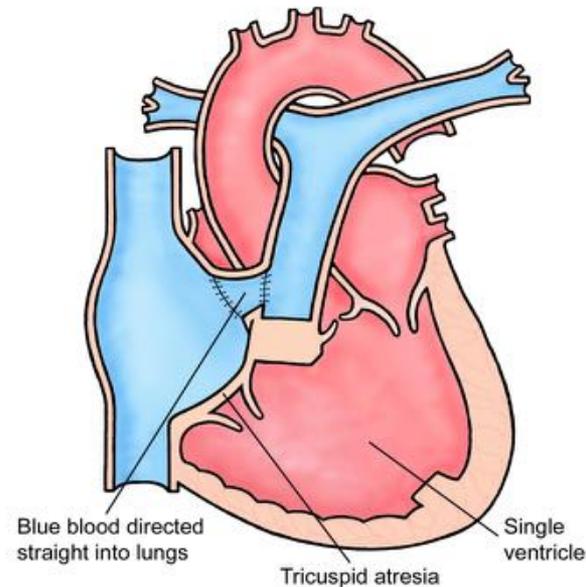
=> Dysfonction sinusale +/- trouble
conductif

Maladie de l'oreillette post opératoire

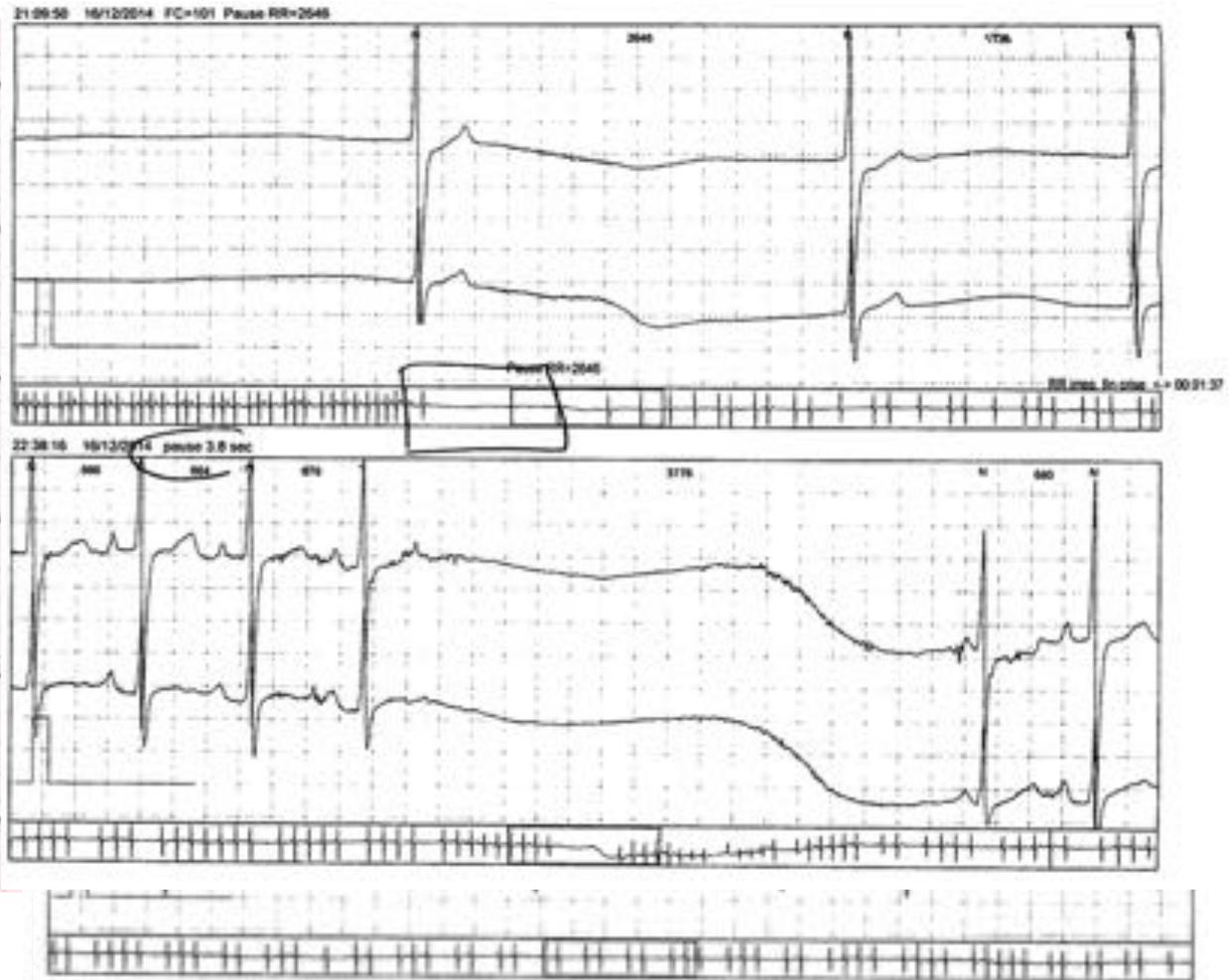
- Mustard/Senning

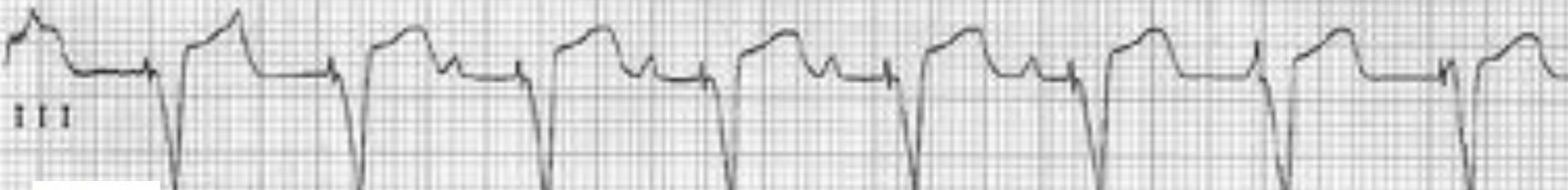
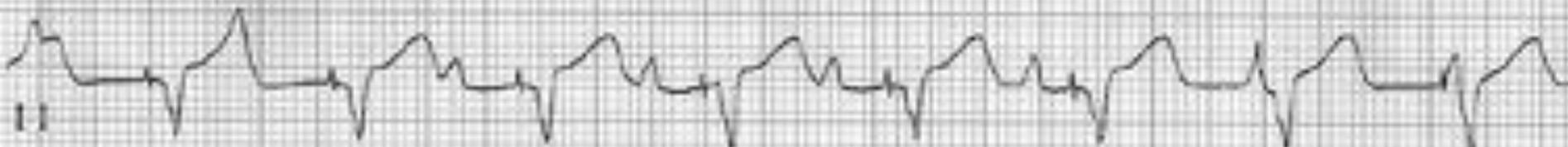
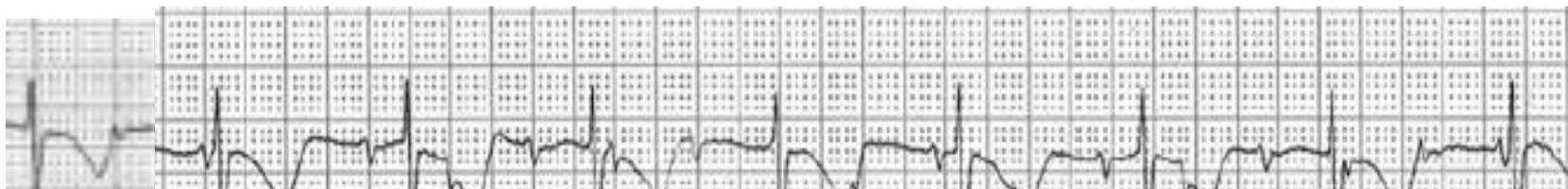


- Fontan/DCPT



Maladie de l'oreillette/Canalopathie





Class I

Sustained pause-dependent VT, with or without prolonged QT, in which the efficacy of pacing is thoroughly documented. (Level of evidence: B)



Class IIa

Long QT syndrome with 2:1 AV or third-degree AV block. (Level of evidence: B)

Indications de stimulation

Dysfonction sinusale et maladie rythmique de l'oreillette

- Class I
 - SYMPTOMES lié à une bradycardie inadaptée à l'âge.

- Class II

- Ttt anti-arythmique pour maladie de l'oreillette (hors digoxine ou RF)

Class IIa

- Cardiopathie et altération hémodynamique / perte de la systole auriculaire

Class IIa

- Bradycardie sévère asymptomatique associée à une cardiopathie congénitale:

Fc < 40 bpm et pauses > 3 sec.

Class IIa pour l'enfant

Class IIb pour l'ado.

- Class III

- Bradycardie asymptomatique: FC > 40bpm et pause < 3 sec.

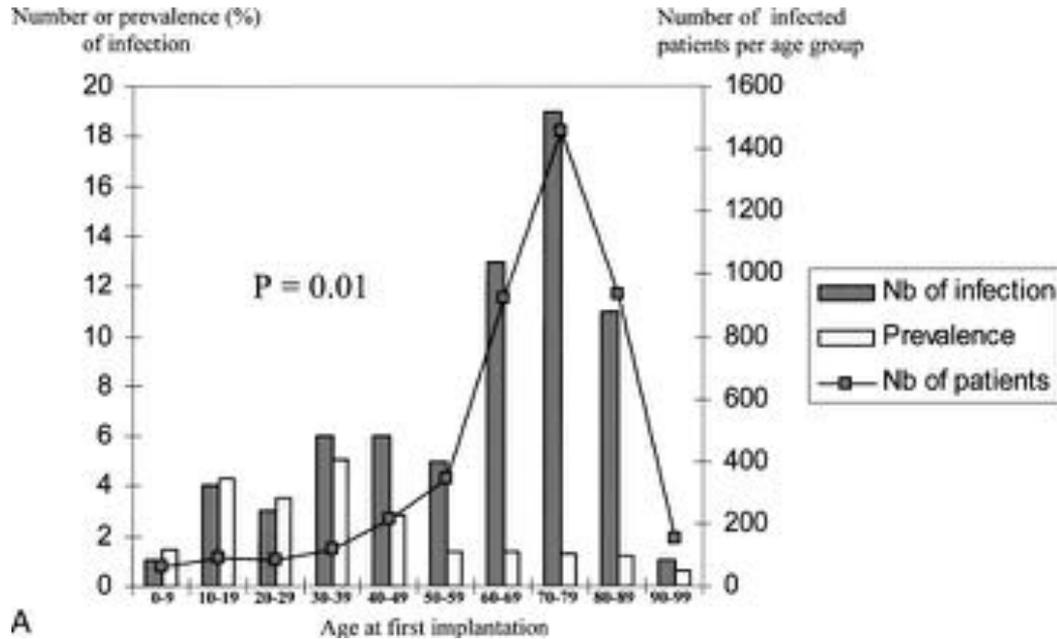
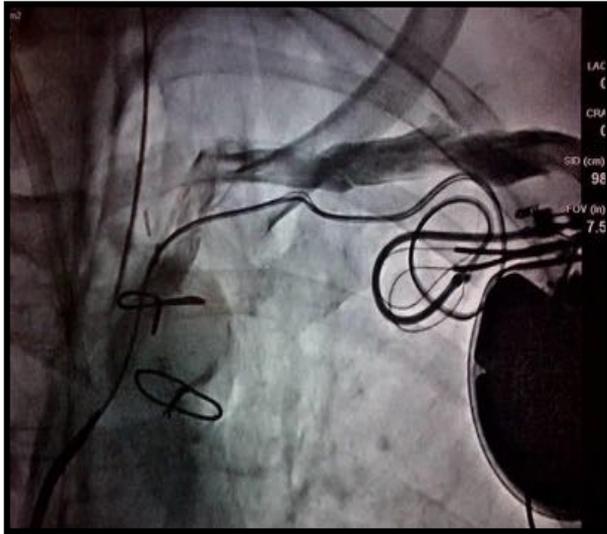
Table 3 Sinus node dysfunction diagnostic criteria⁶

1. Sinus bradycardia
 - Neonates and infants: <60 beats/min asleep and <80 beats/min awake
 - Children aged 2–6 years: <60 beats/min
 - Children aged 7–11 years: <45 beats/min
 - Adolescents and young adults: <40 beats/min
 - Endurance and other highly trained athletes: <30 beats/min
2. Severe sinus arrhythmia (variation in RR interval of ≥100%)
3. Sinus pause or arrest
4. Escape rhythms at slow rate
5. Sinus exit block (second degree, type I and II)
6. Bradycardia/tachyarrhythmia

From *Pediatric arrhythmias: electrophysiology and pacing*
Gillette PC, Garson A

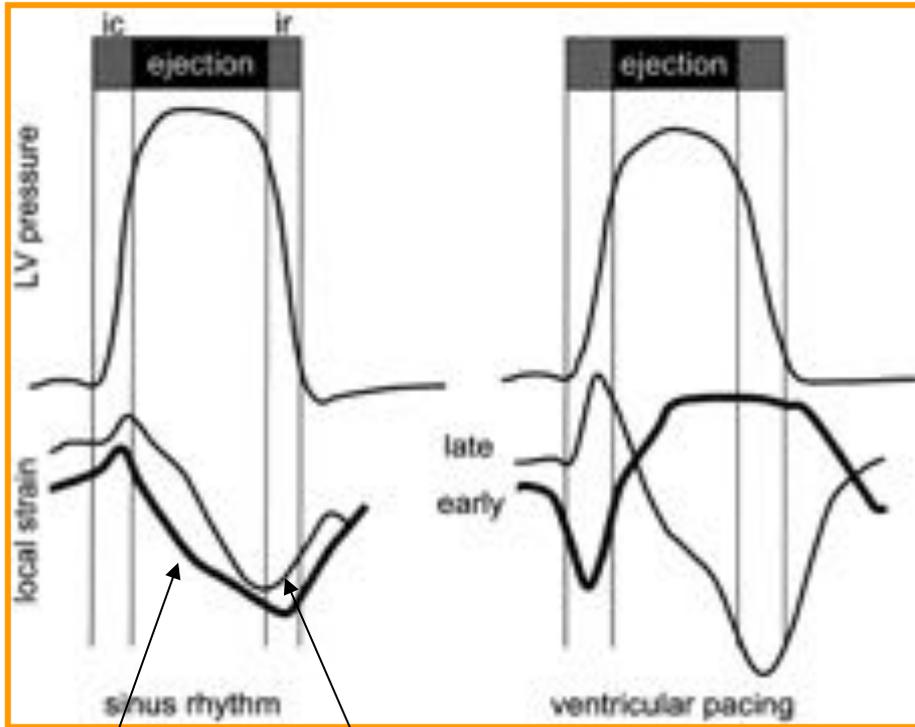
Techniques d'implantation et programmation

Les limites de la stimulation endocardique



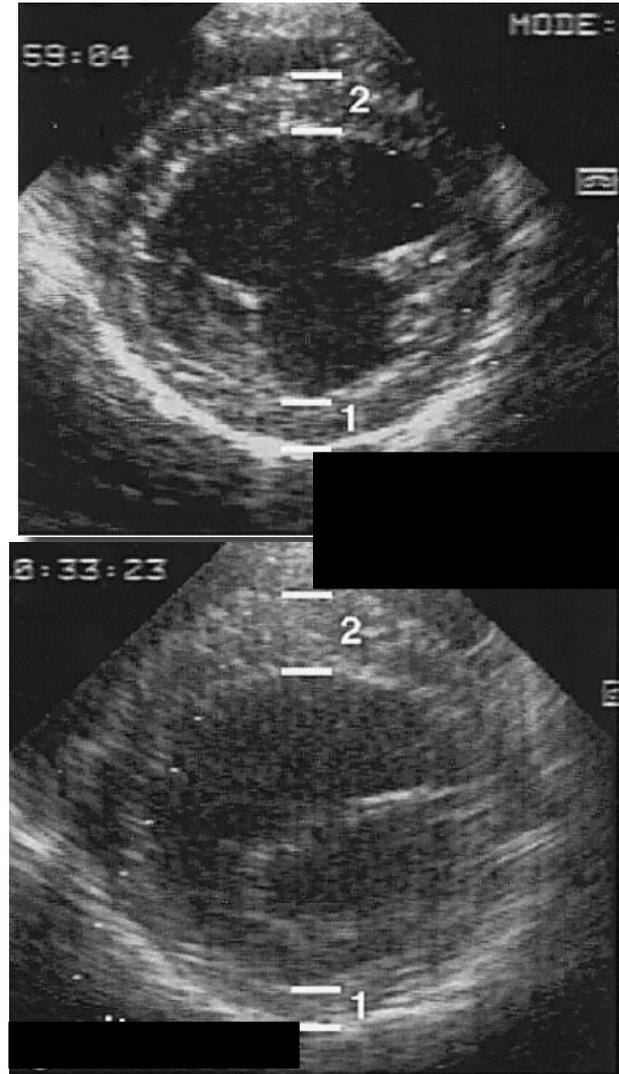
of life

Les limites de la stimulation VD

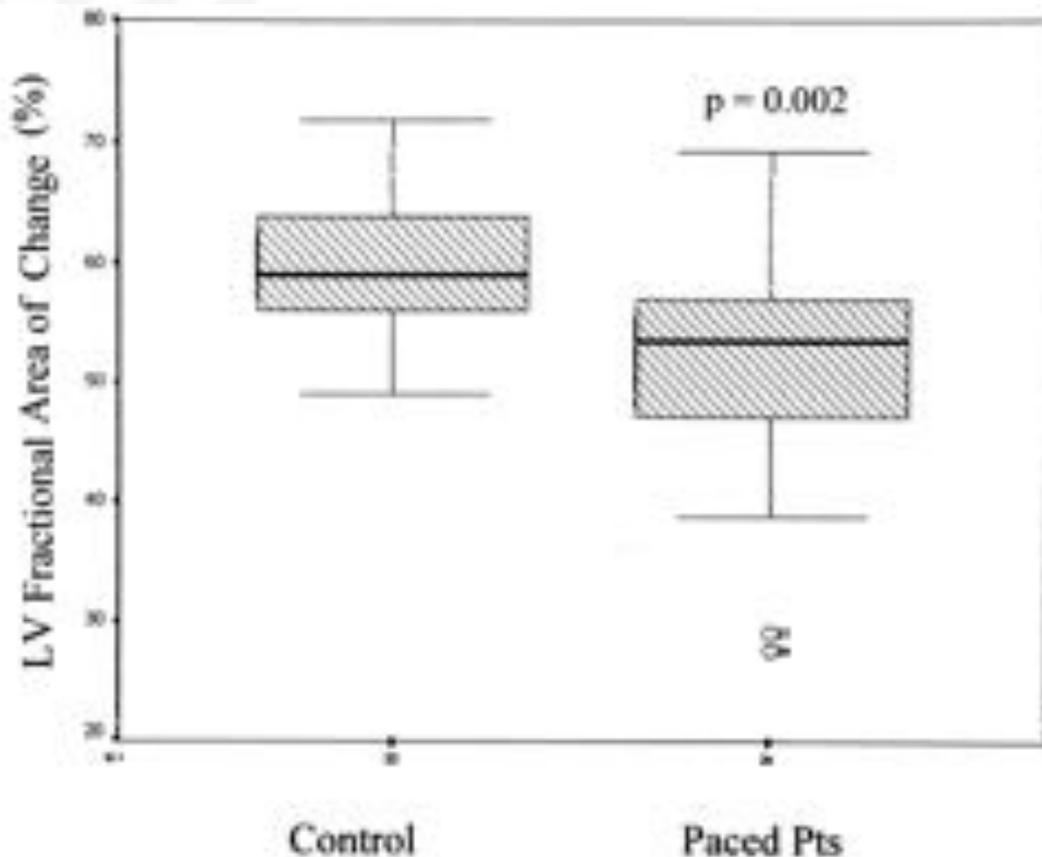


Septal wall

Lateral wall



Detrimental effect of long term apical right ventricular pacing



Long term RVP from the apex appears to be associated with LV systolic and diastolic dysfunction

Tatengco et al JACC 2001

Effet délétère de la sti. VD

REVIEW

The Deleterious Consequences of Right Ventricular Apical Pacing: Time to Seek Alternate Site Pacing

ANTONIS S. MANOLIS

Trial	No. of Patients	Mean Age (y)	Mean FU (y)	LA Diameter	LV Function	CHF	AF
Tantengco et al. ²⁸	24	19.5	9.5	NA	↓	2 pts	NA
Karpawich et al. ²⁹	14	15.5	5.5	NA	Altered Histology	NA	NA
Thambo et al. ³⁰	23	24	10	NA	↓/DS	NA	NA
Tse et al. ³¹	12	72	1.5	NA	↓/MPD	NA	NA
Hamdan et al. ³²	13	66	NA*	NA	↓/↑SNA	NA	NA
DAVID ³⁶	506	64	1	NA	NA	↑	NA
MADIT II ^{37,38} Substudy	567	64	1.7	NA	NA	↑	NA
Wonisch et al. ³⁹	17	59	0.25	NA	NA	**	NA
Thackray et al. ⁴⁰	307	72	5.2	NA	NA		
MOST ⁴¹	1,339	74	6	NA	NA		
Nielsen et al. ⁴³	177	74	2.9	↑	↓		
O'Keefe et al. ⁴⁴	59	69	1.5	NA	↓		

AF = atrial fibrillation; CHF = congestive heart failure; DS = dyssynchrony; FU = follow-up; LA = left atrium; LBBB = left bundle branch block; LV = left ventricular; MPD = myocardial perfusion defects; NA = not available/ not assessed; SNA = sympathetic activity

*Acute study.
**Permanent RV pacing significantly reduced exercise capacity and submaximal cardiorespiratory parameters.

Detrimental Ventricular Remodeling in Patients With Congenital Complete Heart Block and Chronic Right Ventricular Apical Pacing

Jean-Benoît Thambo, MD*; Pierre Bordachar, MD*; Stéphane Garrigue, MD, PhD; Stéphane Lafitte, MD, PhD; Prashanthan Sanders, MBBS, PhD; Sylvain Reuter, MD; Romain Girardot, MD; David Crepin, MD; Patricia Reant, MD; Raymond Roudaut, MD; Pierre Jaïs, MD; Michel Haïssaguerre, MD; Jacques Clementy, MD; Maria Jimenez, MD, PhD

LIRYC | Restoring the rhythm

TABLE 2. Comparison Between Controls and Patients After Long-Term Follow-Up

	Long-Term RV Pacing	Controls
Cardiac output, L/min	3.8±0.6*	4.9±0.8
Mean LV EDD, mm	55±7*	46±6
Pathological LV EDD, %	52†	0
Ratio posterior/septal wall	1.3±0.2†	1±0.1
Ratio mitral regurgitation/left atrium	16±8*	5±2
LV filling time, ms	415±39*	477±51
Interventricular dyssynchrony, ms	55±18†	18±11
Intra-LV delay, ms	59±18†	19±9
Septal/posterior wall delay, ms	84±26†	18±9
DLC, %	39±15†	10±7
Exercise, W	123±24†	185±39

EDD indicates end-diastolic diameter.

*P<0.05; †P<0.01.

Predictors of left ventricular remodelling and failure in right ventricular pacing in the young

Roman A. Gebauer¹, Viktor Tomek¹, Aida Salameh², Jan Marek³,
Václav Chaloupecký¹, Roman Gebauer¹, Tomáš Matějka¹,
Pavel Vojtovič¹, and Jan Janoušek^{2*}

¹Noninvasive and Cardiovascular Research Center, University Hospital Masch, Brno, Czech Republic; ²Department of Pediatric Cardiology, University of Leipzig Heart Center, Semmelweisstr. 25, 04089 Leipzig, Germany; and ³Department of Pediatric Cardiology, Great Ormond Street Hospital, London, UK

Received 17 May 2007, revised 11 December 2008, accepted 10 January 2009, online published ahead of print 12 March 2009

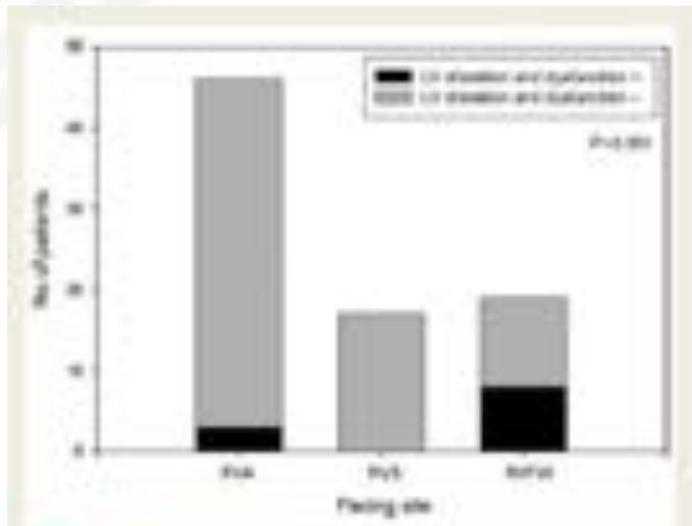


Figure 1 Proportion of patients developing LV dilatation and dysfunction (SF < 0.26 and LVEDD > +2z-values) according to the pacing site. RVA, endocardial RV apical pacing; RVF, endocardial RV septal pacing; RVFW, epicardial RV free wall pacing

Aims

To identify risk factors for left ventricular (LV) dysfunction in right ventricular (RV) pacing in the young.

Methods and results

Left ventricular function was evaluated in 82 paediatric patients with either non-surgical ($n = 41$) or surgical ($n = 41$) complete atrioventricular block who have been 100% RV paced for a mean period of 7.4 years. Left ventricular shortening fraction (SF) decreased from a median (range) of 39 (24–62)% prior to implantation to 32 (8–49)% at last follow-up ($P < 0.05$). Prevalence of a combination of LV dilatation (LV end-diastolic diameter > +2z-values) and dysfunction (SF < 0.26) was found to increase from 1.3% prior to pacemaker implantation to 13.4% (11/82 patients) at last follow-up ($P = 0.01$). Ten of these 11 patients had progressive LV remodelling and 8 of 11 were symptomatic. The only significant risk factor for the development of LV dilatation and dysfunction was the presence of epicardial RV free wall pacing (OR = 14.3, $P < 0.001$). Other pre-implantation demographic, diagnostic, and haemodynamic factors including block aetiology, pacing variables, and pacing duration did not show independent significance.

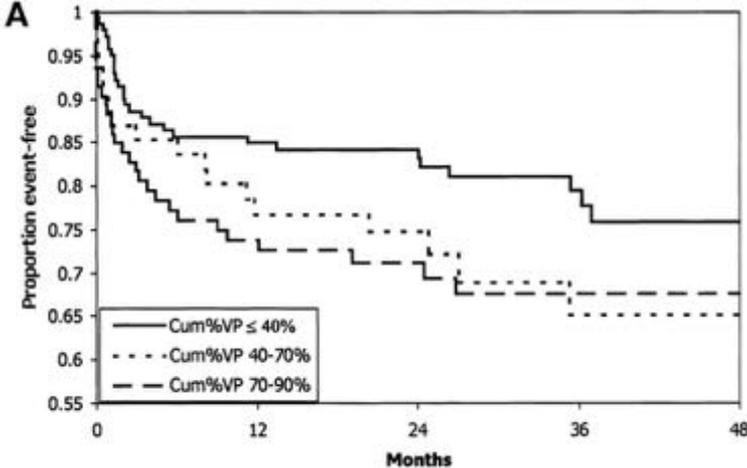
Conclusion

Right ventricular pacing leads to pathologic LV remodelling in a significant proportion of paediatric patients. The major independent risk factor is the presence of epicardial RV free wall pacing, which should be avoided whenever possible.

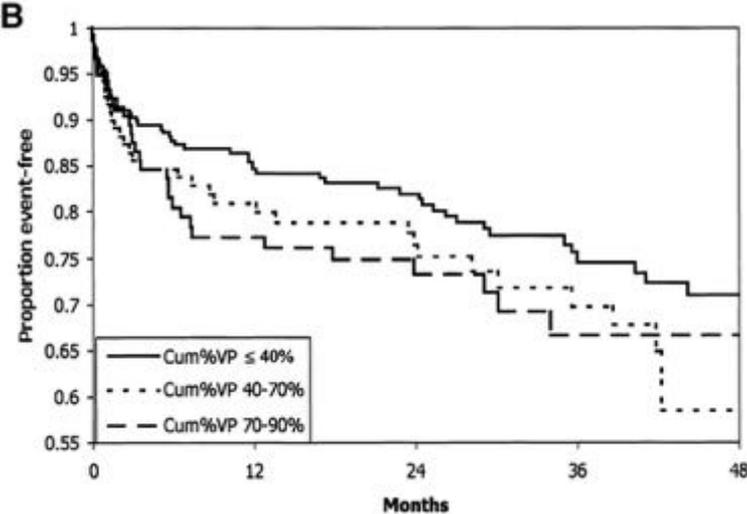
Selecting pacing sites in children with complete heart block: is it time to avoid the right ventricular free wall?

Luc Mertens* and Mark K. Friedberg

Les limites de la stimulation VD



Atrial fibrillation



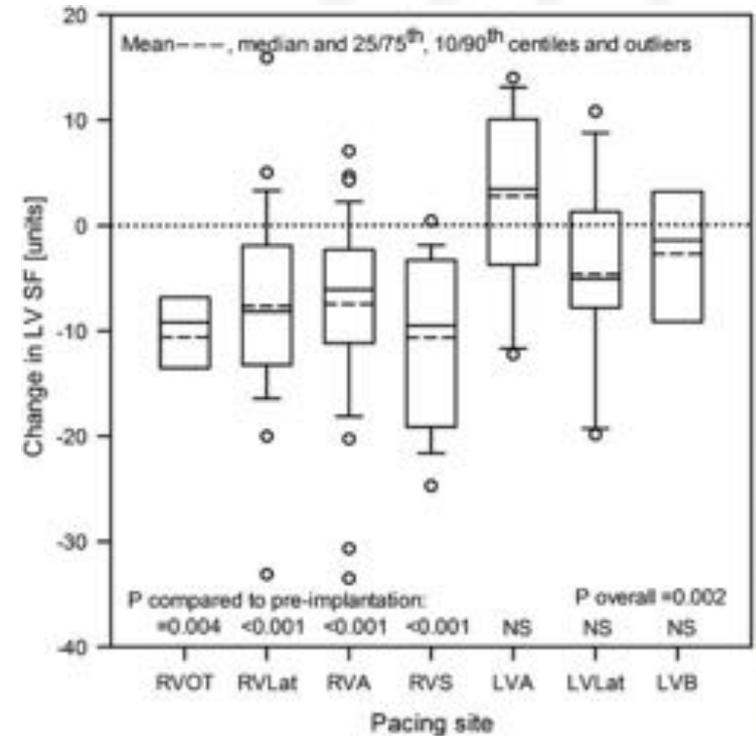
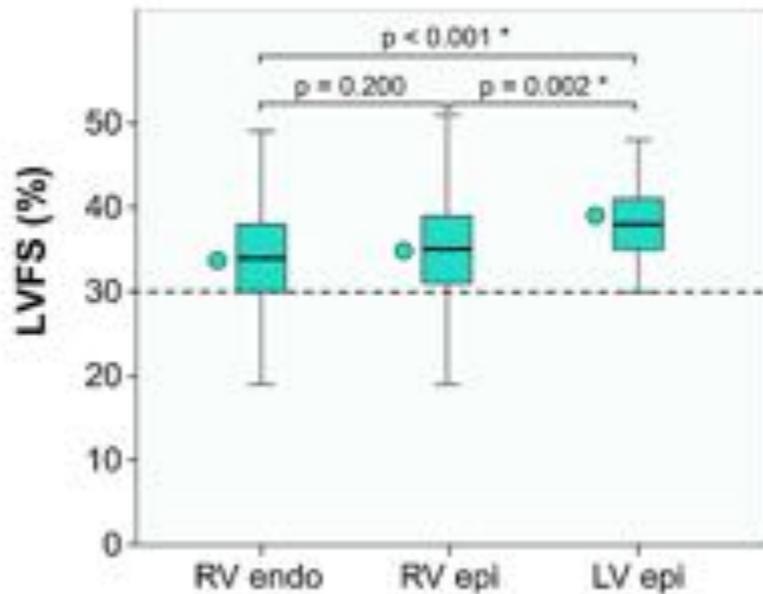
Kaplan-Meier rates for freedom from first documented incidence of atrial fibrillation

Best site to pace ?

La stimulation ventriculaire droite permanente expose à un remodelage ventriculaire délétère

Particularités de la population pédiatrique:

- Stimulés toute leur vie
- Stimulés pendant la croissance cardiaque



Pacing site and configuration

< 15 KG	Epicardial	LV apex or lat
15-25 KG	Epicardial	RA/LV LV
> 25 KG	Epicardial	RA/LV LA/LV
30-40 ans	Epicardial	BIV or LV

100 % chirurgical

Stimulation multi-sites et resynchronisation en pédiatrie

Table 10.1. Summary of clinical studies evaluating CRT in CHD

Author	Year	No. patients	Age (years)	CHD%	Systemic RV%	Single V%	Coro pacing%	NYHA III-IV%	QRS ms	EF pre%	EF post%	Nonresp %	Design and main features
Janosik et al	2004	8	15.0 [†] (6.9-29.2)	100	100	0	75.0	12.5	164 [‡]	35 [‡]	30 [‡]	—	Single-center, prospective, first study on utility of CRT in systemic right ventricle
Debin et al	2005	103	12.8 [†] (0.3-55.4)	70.9	36.5	6.8	44.7	37.9	166 [*]	26 [*]	40 [*]	10.7	Multicenter, retrospective, first large study on CRT in congenital heart disease
Khairy et al	2006	13	7.8 [*] (0.8-15.5)	100	30.8	0	100	—	>120 in all	31 [*]	51 [*]	11.1	Single-center, retrospective, impaired ventricular function and conduction abnormality in all, follow-up 17 months
Musk et al	2006	6	11.3 [*] (0.5-23.7)	33.3	0	0	100	—	204 [*]	36 [*]	60 [*]	0.0	Single-center, retrospective, super-response after upgrade from conventional right ventricular pacing to CRT
Grubb et al	2009	60	15.0 [†] (0.5-47.0)	76.7	15.0	21.7	68.3	31.7	100 [‡]	36 [‡]	42 [‡]	10.0	Single-center, retrospective, largest reported single ventricular patient group
Jacout et al	2009	7	24.6 [*] (15.0-50.0)	100	100	0	71.4	100.0	100 [*]	—	—	—	Single-center, prospective, effect of CRT in systemic systemic right ventricle
Janosik et al	2009	109	16.9 (0.3-73.8)	79.8	33.0	3.7	77.1	45.9	100 [‡]	30 [*]	41 [*]	13.7	Multicenter, retrospective, effects of CRT in different structural and functional substrates
Thambis et al	2013	9	36.6 [*] (>10)	100	0	0	0	—	166 [*]	50 [*]	56 [*]	—	Single-center, prospective, postoperative strategy of failed, noninvasive mapping of ventricular activation

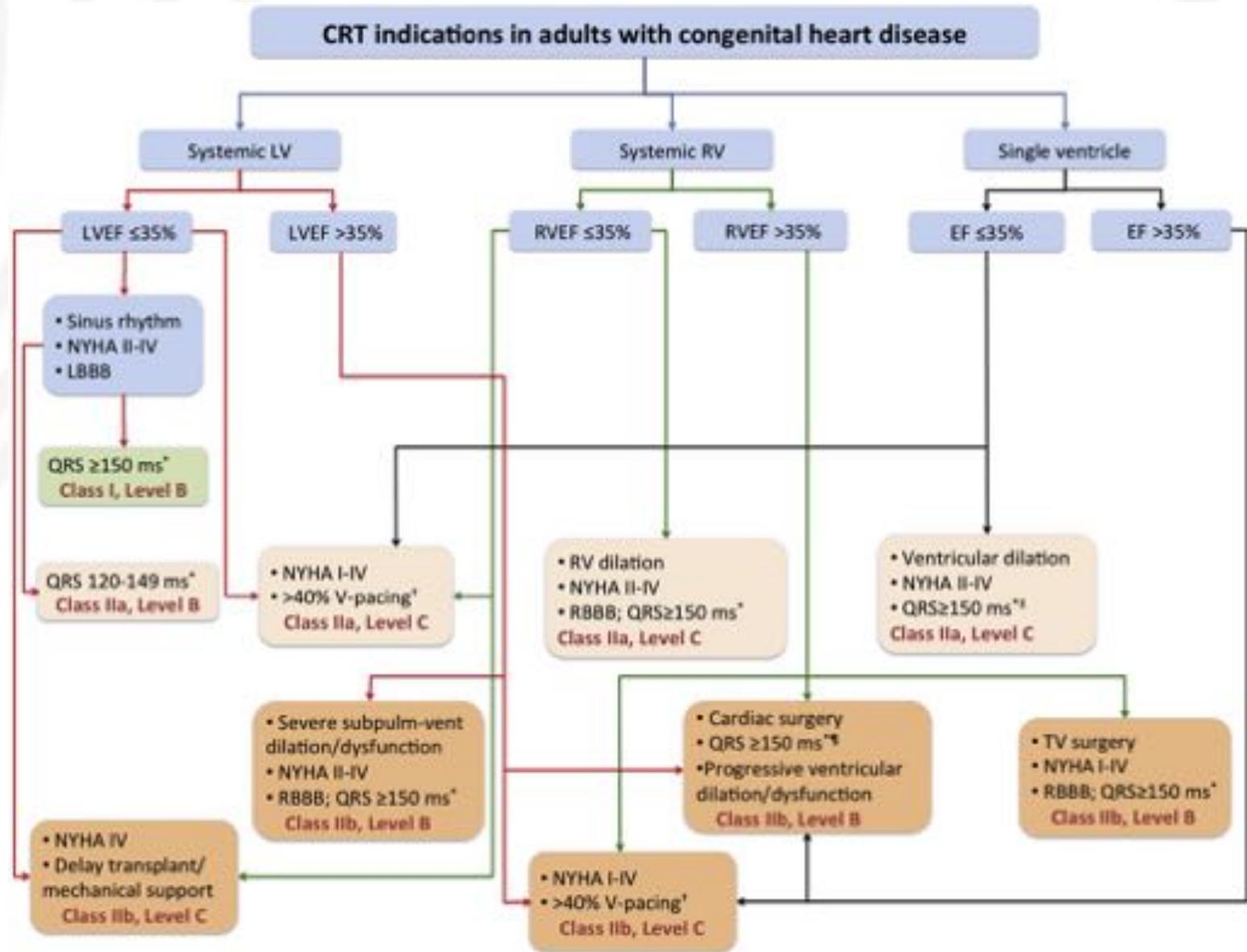
CHD = congenital heart disease; Coro pacing = conventional pacing prior to cardiac resynchronization therapy (CRT); EF post = ejection fraction following CRT; EF pre = ejection fraction prior to CRT; Nonresp = nonresponder; NYHA = New York Heart Association; RV = right ventricle; Single V = single ventricle.

* Mean value.

† Median value.

‡ Right ventricular fractional area of change.

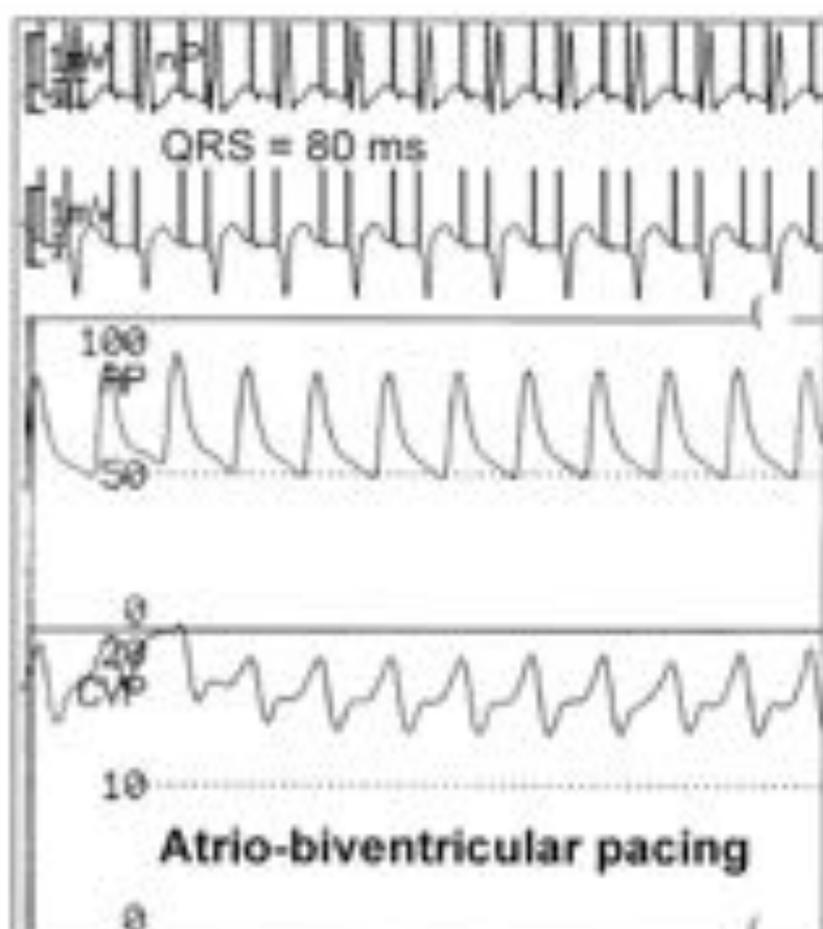
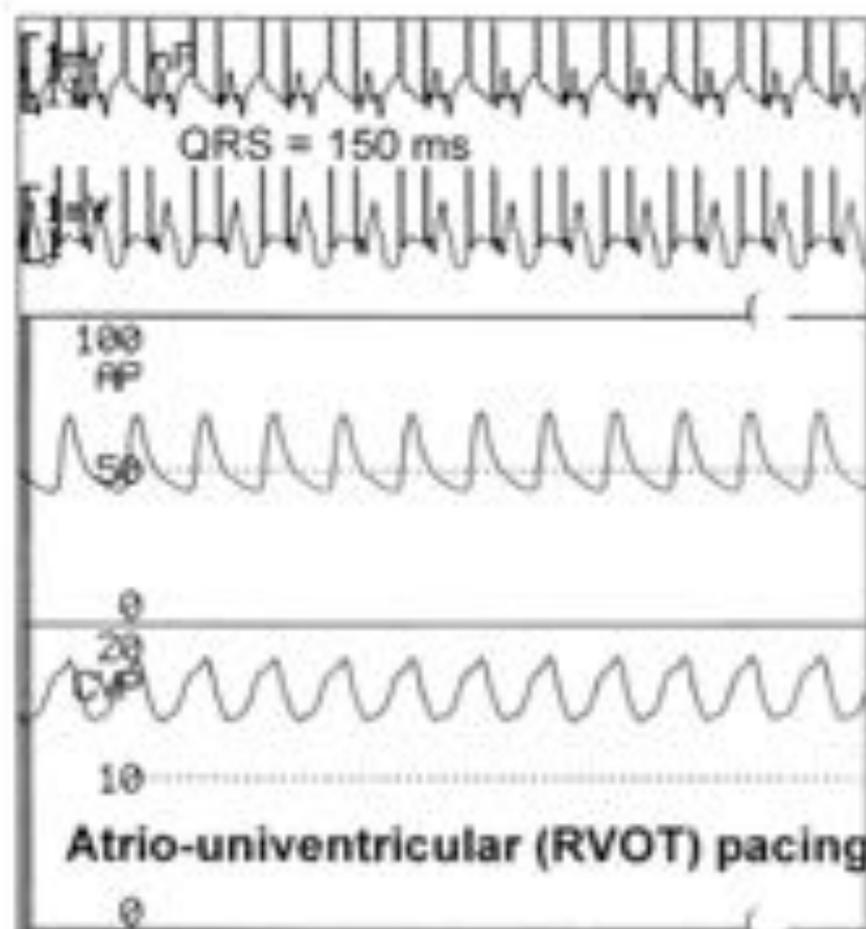
Khairy et al; PACES/HRS Expert Consensus Statement on Arrhythmias in Adult Congenital Heart Disease 2014



Khairy et al, Canadian Journal of Cardiology, 2014

Resynchronization Pacing Is a Useful Adjunct to the Management of Acute Heart Failure After Surgery for Congenital Heart Defects

Jan Janoušek, MD, Pavel Vojtovič, MD, Bohumil Hučín, MD, Tomáš Tláškal, MD, Roman Antonín Gebauer, MD, Roman Gebauer, MD, Tomáš Matějka, MD, Jan Marek, MD, and Oleg Reich, MD

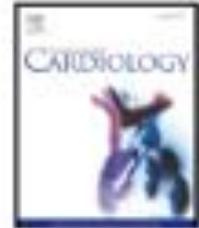




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Letter to the Editor

“De novo” biventricular pacing in two children with complete atrio-ventricular block and severe ventricular dilatation: Early reverse remodeling ☆

Antonio Ammirati, Massimo Stefano Silveti*, Duccio Di Carlo, Fabio Anselmo Saputo, Antonio Longoni, Fabrizio Drago



1 week later

LIRYC | Restoring the rhythm of life

Acute Hemodynamic Benefit of Left Ventricular Apex Pacing in Children

Ward Y. Vanagt, MD, Xander A. Verbeek, PhD, Tammo Delhaas, MD, PhD, Marc Gewillig, MD, PhD, Luc Mertens, MD, PhD, Patrick Wouters, MD, PhD, Bart Meyns, MD, PhD, Willem J. Daenen, MD, and Frits W. Prinzen, PhD

Departments of Physiology and Pediatrics, Cardiovascular Research Institute Maastricht, Maastricht, The Netherlands, and Departments of Pediatric Cardiology, Anesthesiology, and Cardiothoracic Surgery, University Hospital Gasthuisberg, Catholic University of Leuven, Leuven, Belgium

Background. Despite the fact that pacing at the right ventricular apex acutely and chronically decreases left ventricular contractile function, this pacing site is still conventionally used in adults and children. Because animal studies showed beneficial effects of left ventricular pacing, we compared the hemodynamic performance of left ventricular apex, left ventricular free wall, and right ventricular apex pacing in children.

Methods. Studies were performed in 10 children (median age, 2.5 years; range, 2 months to 17 years) undergoing surgery for congenital heart disease with normal systemic left ventricular anatomy and intraventricular conduction. High-fidelity left ventricular and arterial pressure measurements were performed during epicardial right ventricular apex and left ventricular apex and free wall pacing.

Results. Left ventricular apex pacing increased the maximum rate of rise of left ventricular pressure and pulse pressure significantly relative to right ventricular apex pacing (by $7.7\% \pm 7.2\%$ and $7.7\% \pm 7.0\%$, respectively) without changes in end-diastolic left ventricular pressure. Left ventricular free wall pacing did not significantly improve hemodynamics as compared with right ventricular apex pacing. The QRS duration was not different among pacing at the three sites.

Conclusions. In this short-term study left ventricular apex pacing is hemodynamically superior to right ventricular apex and left ventricular free wall pacing in children. Therefore, the left ventricular apex appears a favorable pacing site after pediatric cardiac surgery.

(Ann Thorac Surg 2005;79:932-6)

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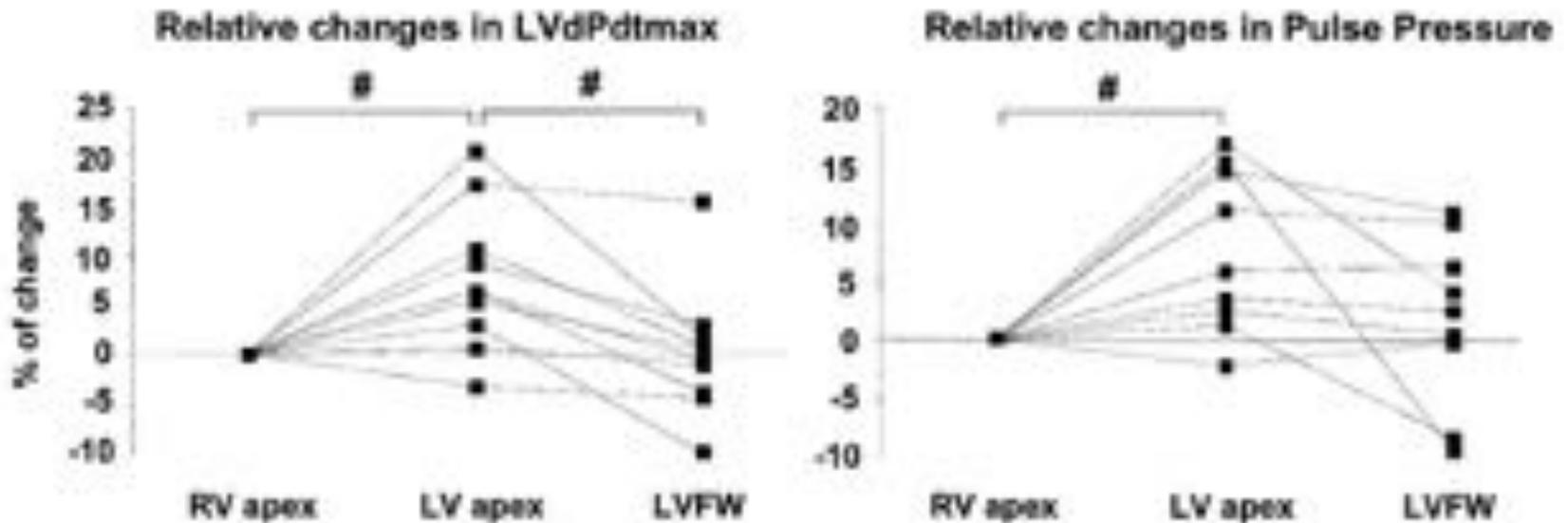


Fig 1. Percentage changes in maximal rate of rise of left ventricular pressure (LVdPdtmax; left panel, $n = 10$) and in pulse pressure (right panel, $n = 9$) for individual patients with right ventricular (RV) apex pacing as reference value (# = $p < 0.05$ versus left ventricular apex pacing). (RV apex = RV apex pacing; LV apex = left ventricular apex pacing; LVFW = left ventricular free wall pacing.)

Un double chambre pour qui ?

Question

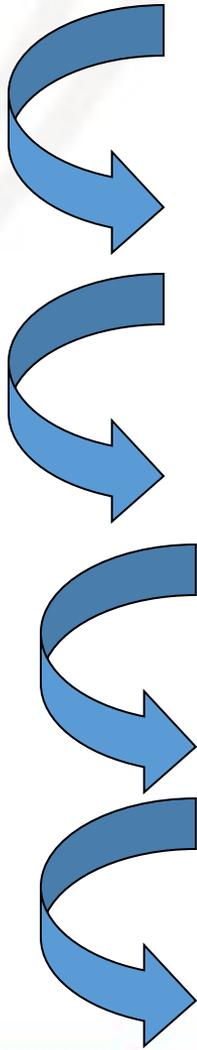
1. Dès 25 kg, il est classique de passer en endocavitaire double chambre
2. Je reste en VVIR le plus longtemps possible car il est préférable d'implanter le moins de sonde possible
3. Les capteur d'asservissement sont aujourd'hui très efficace et évitent les sous détections atriale avec chute brutale de fréquence de stimulation souvent mal tolérés
4. Un simple chambre est classiquement implanté lors de troubles conductifs A-V intermittents

Pacing site and configuration

< 15 KG	Epicardial	LV apex ? or lat
15-25 KG	Epicardial	RA/LV LV
> 25 KG	Epicardial	RA/LV LA/LV
30-40 ans	Epicardial	BIV or LV

100 % chirurgical

Besoin d' une technique alternative



Stimulation plus physiologique

BiV ou VG

Pas de sonde endocavitaire

Le moins invasif possible

Durabilité longue

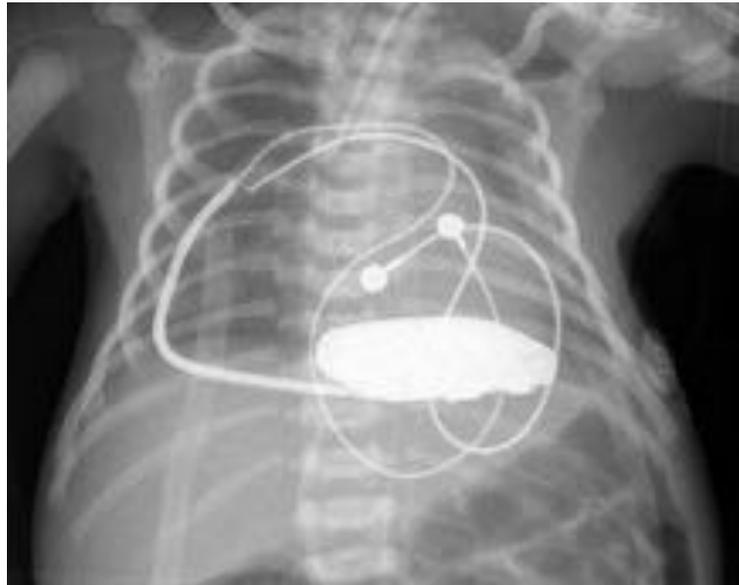
News strategies...

Interactive Cardiovascular and Thoracic Surgery 9 (2009) 743-745

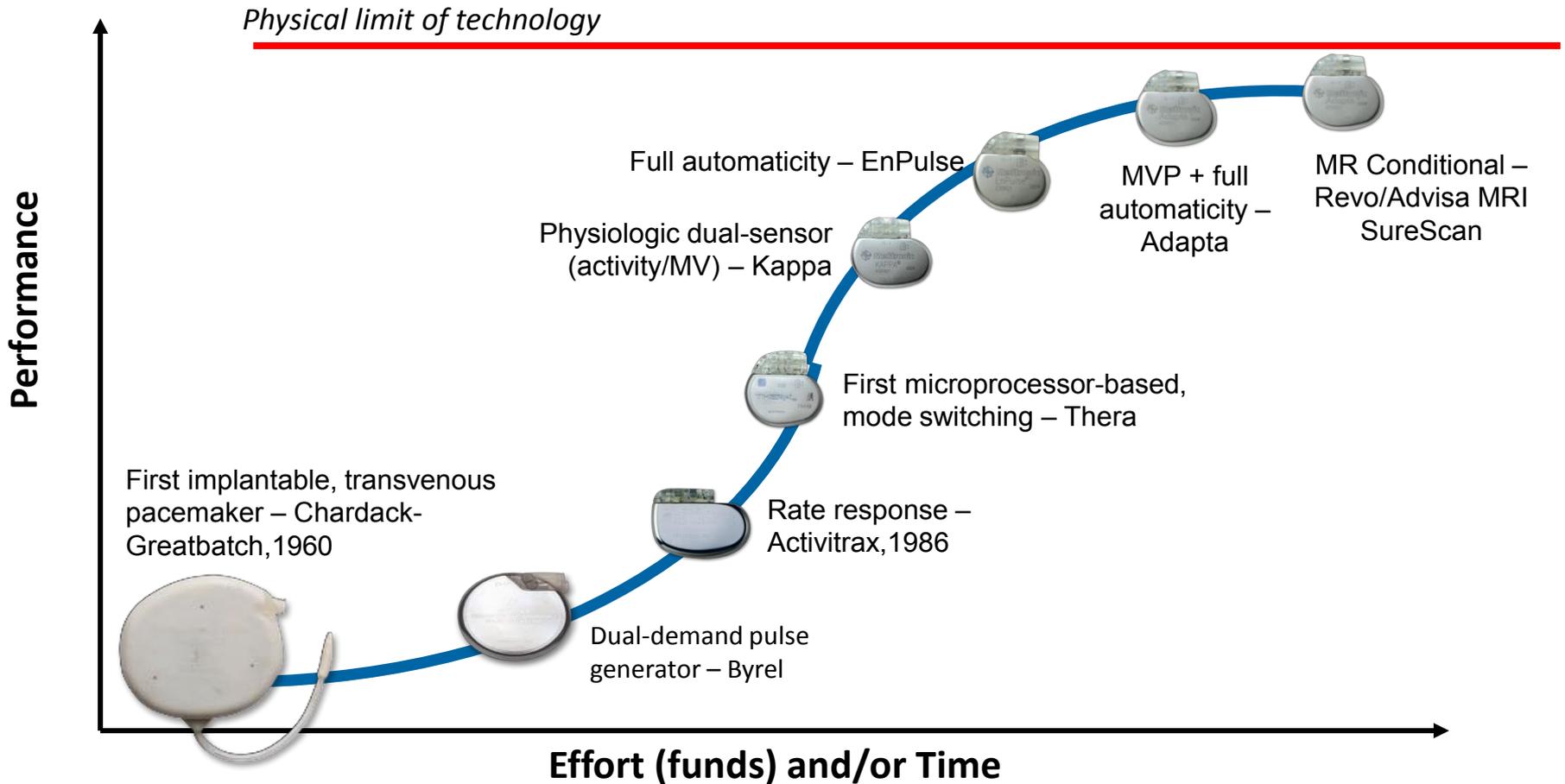
www.icvts.org

Case report - Congenital Intra-diaphragmatic pacemaker implantation in very low weight premature neonate

François Roubertie^a, Emmanuel Le Bret^{b,*}, Jean Benoit Thambo^a, Xavier Roques^a

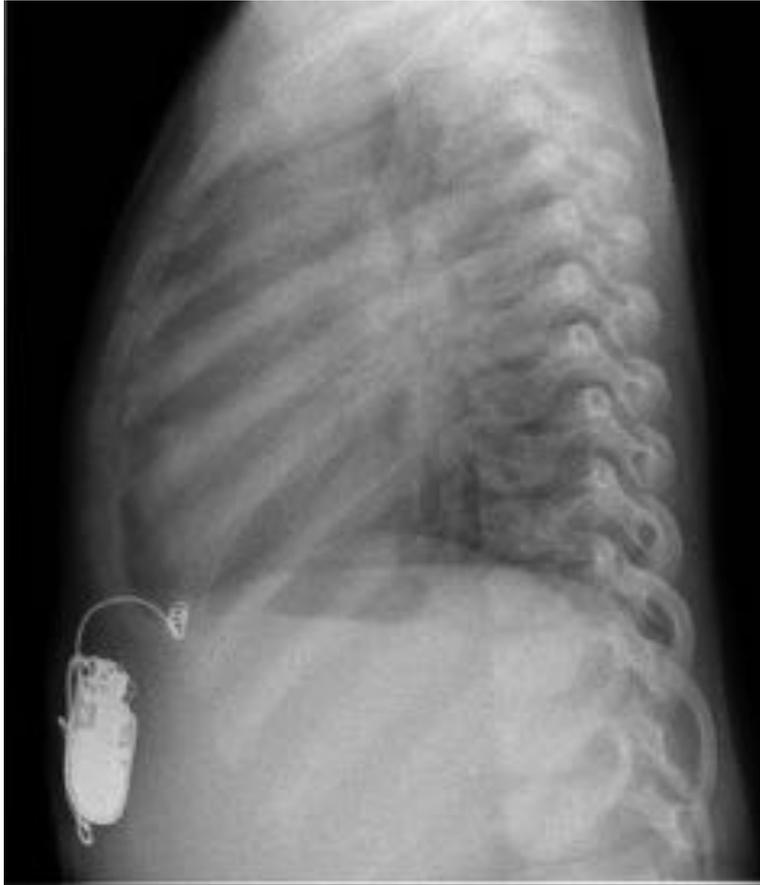


Limites de la stimulation endocardique traditionnelle



Preformance technologique suit une courbe en S

Stimulation épiscopardique chirurgicale



BAV congénital
Sonde VD



BAV congénital + myocardiopathie dilatée
Sonde OD + 2 sondes VG

Dans un monde parfait ...



Stimulation plus physiologique

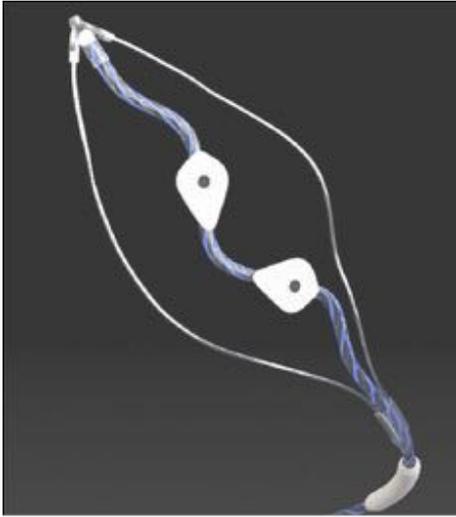


Peu invasif

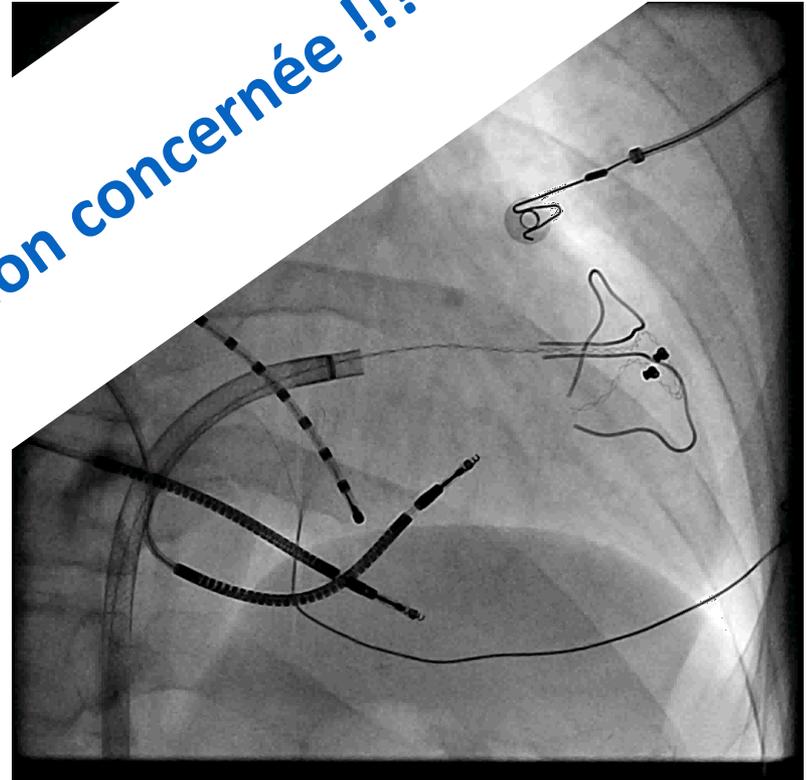


Pas de sonde endocavitaire

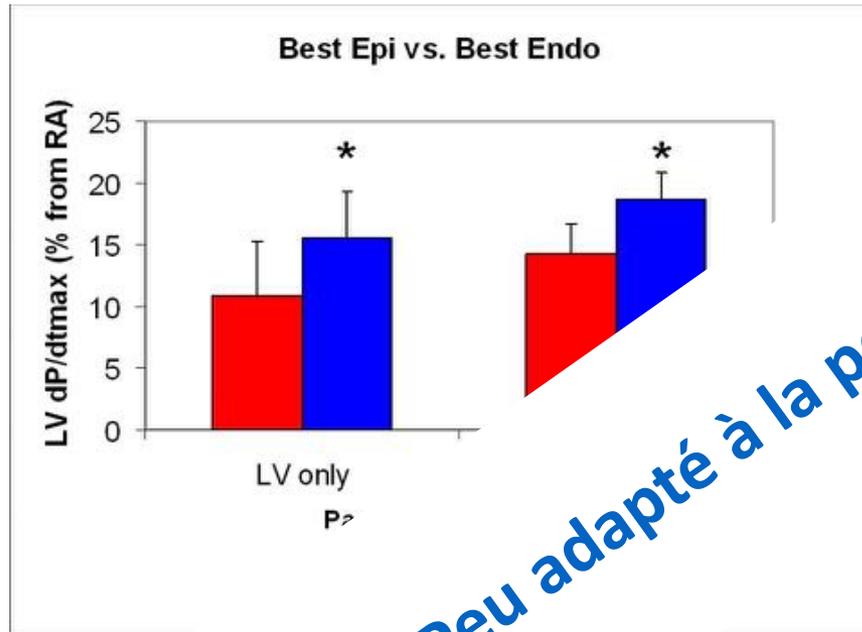
Stimulation épiscopardique souxxyphoidienne



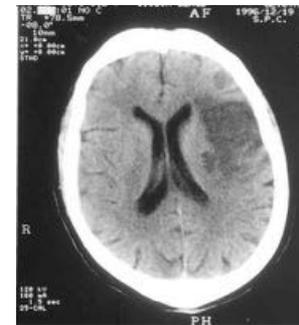
Peu adapté à la population concernée !!!



Stimulation endocardique ventriculaire gauche

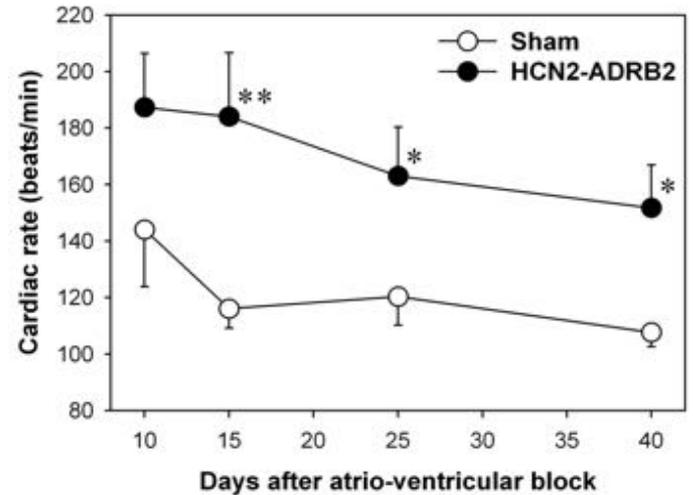
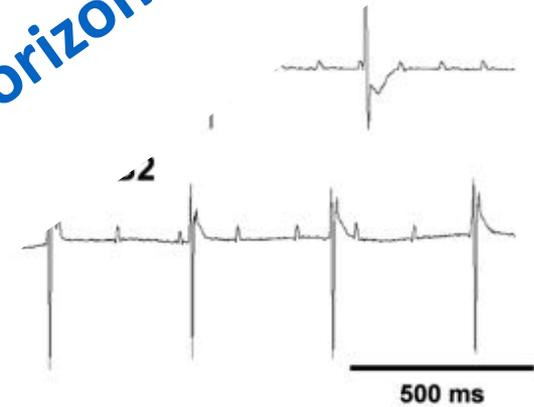
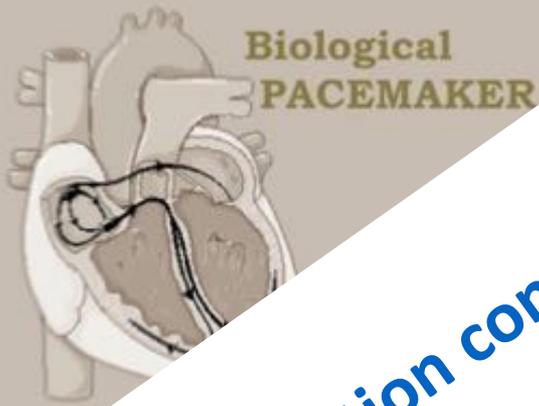


Peu adapté à la population concernée !!!

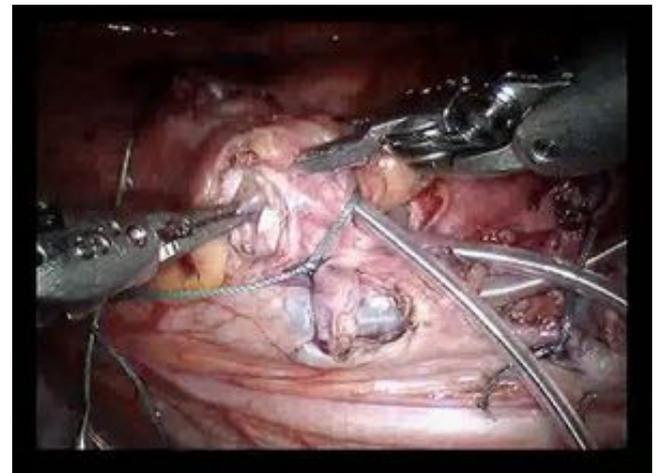


Pacemaker biologique

Adapté à la population concernée mais horizon lointain !!!

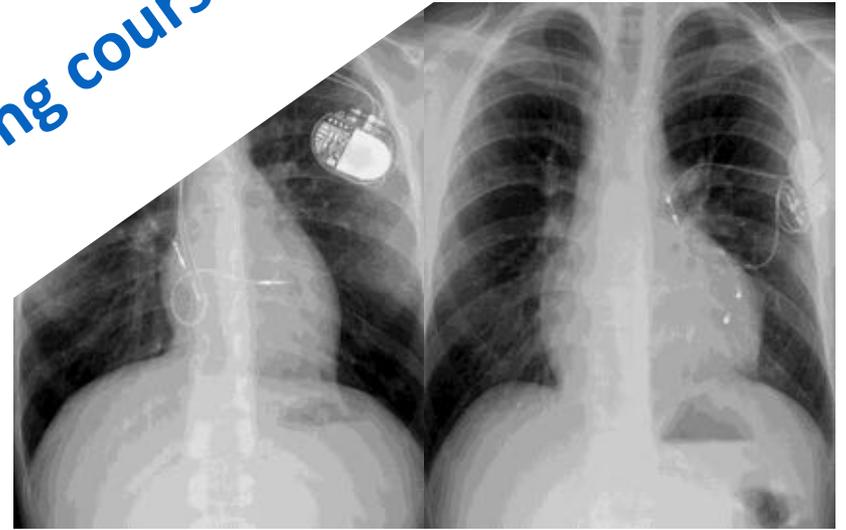
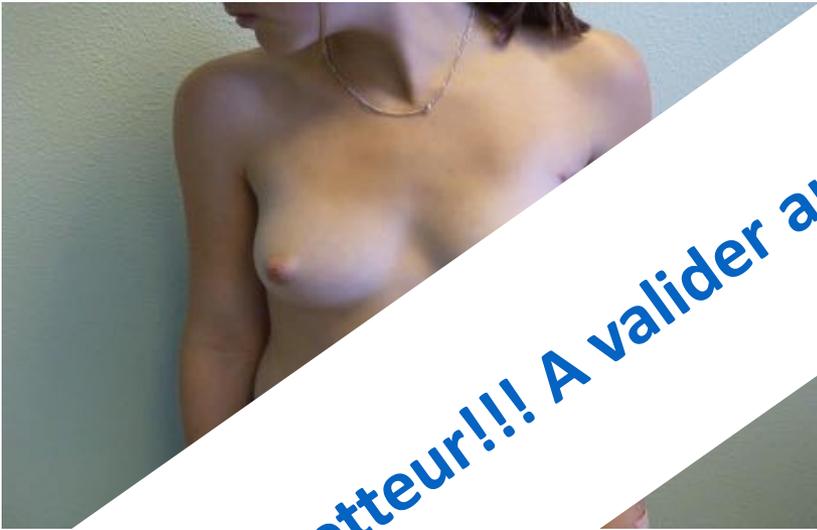


Implantation robotisée: sonde épicardique VG

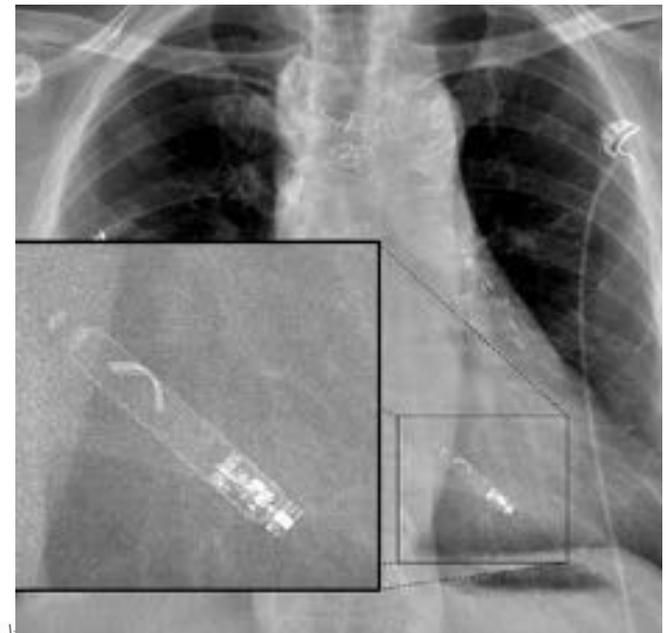
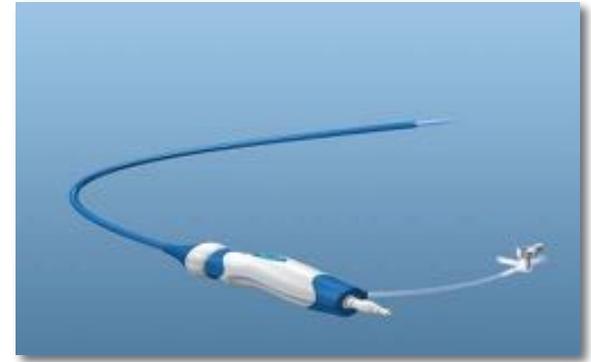


Implantation robotisée: sondes épicardique VG

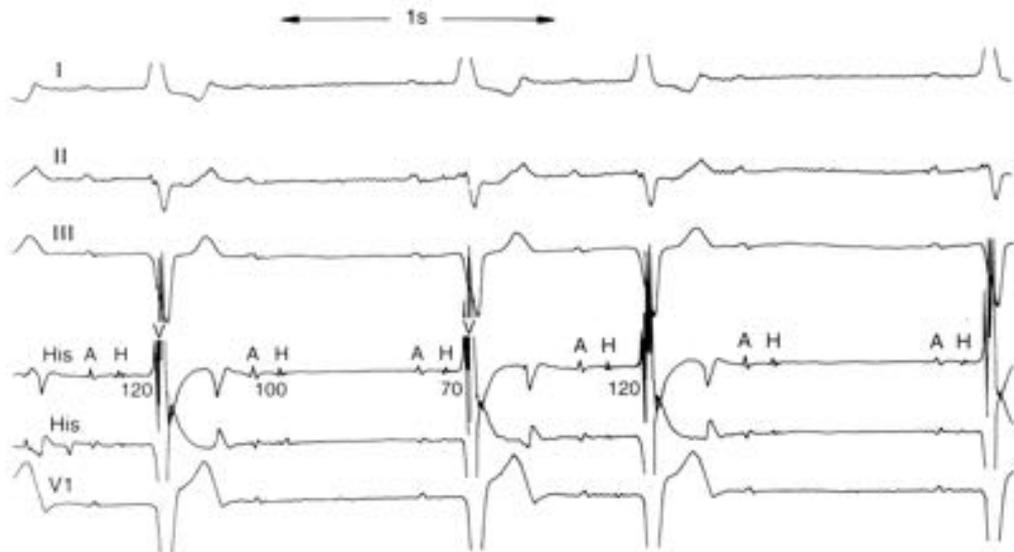
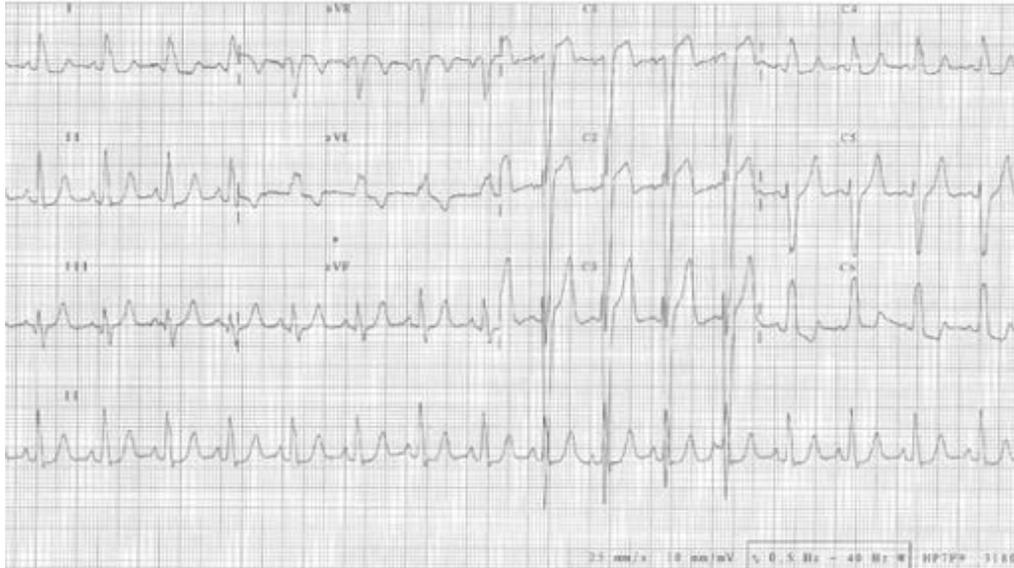
Prometteur!!! A valider au long cours et à large échelle



Stimulateur sans sonde



Patient 69 ans; syncope



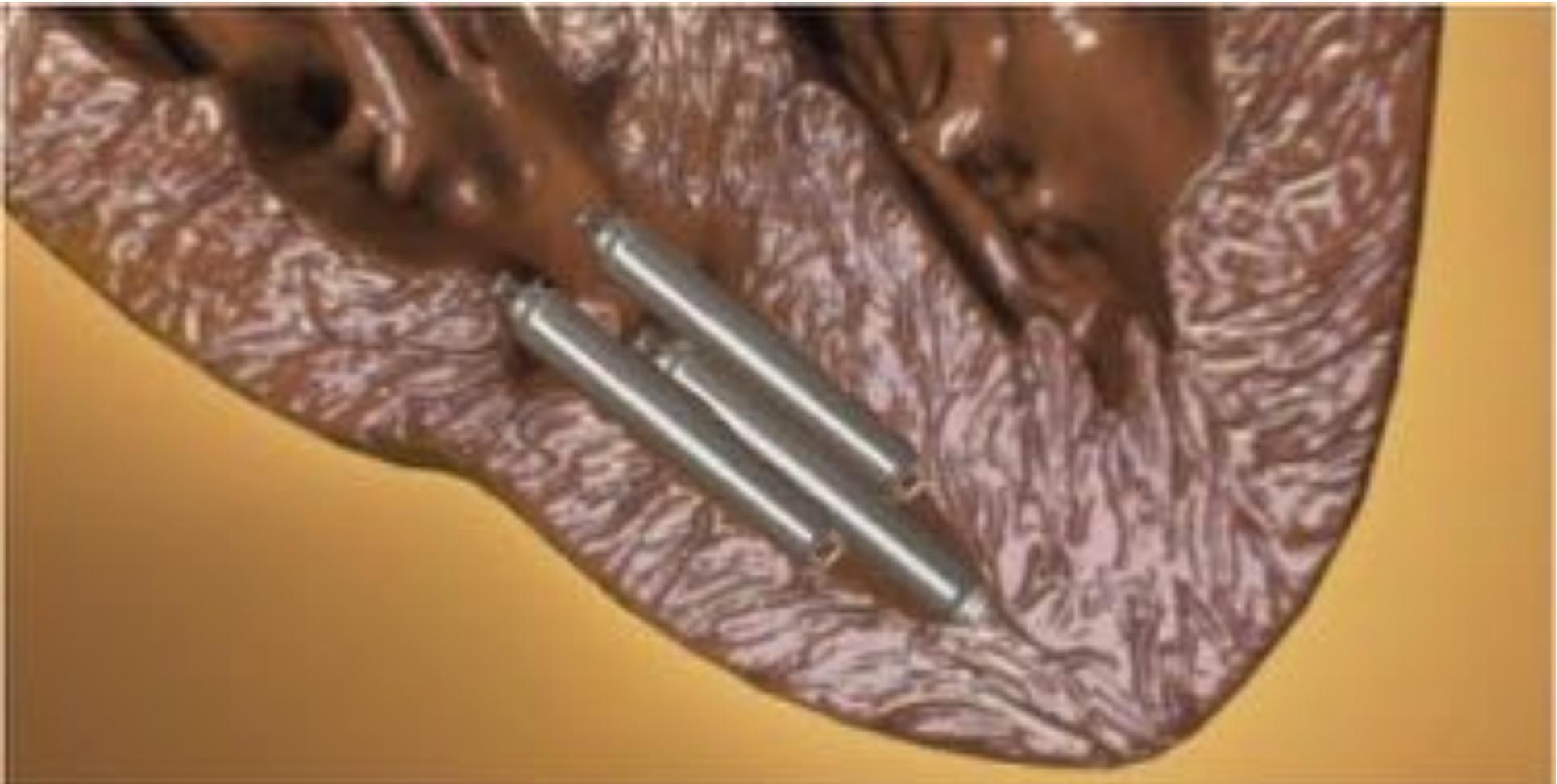
Les limites: que faire en fin de vie du dispositif ???



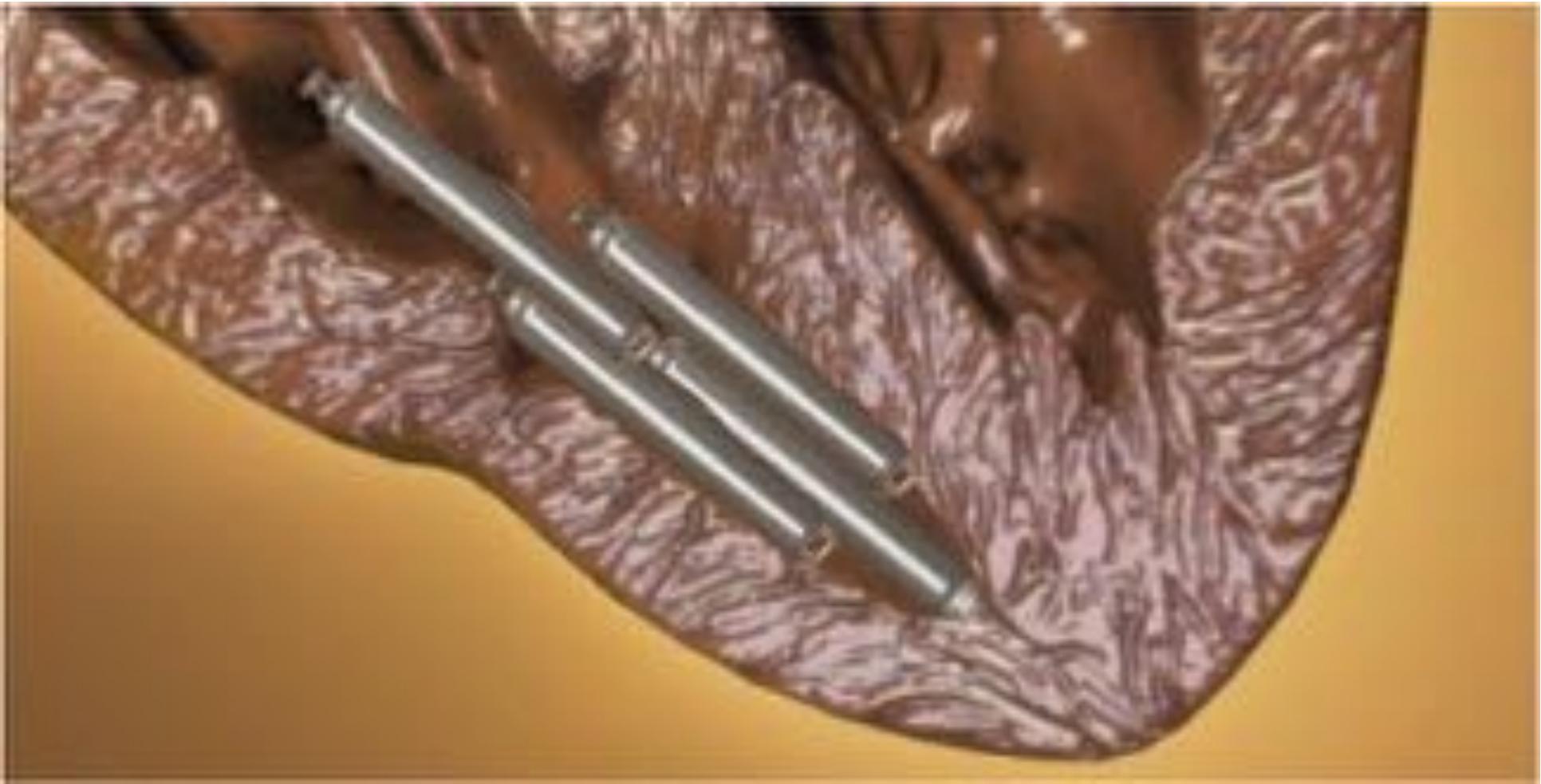
Les limites: que faire en fin de vie du dispositif ???



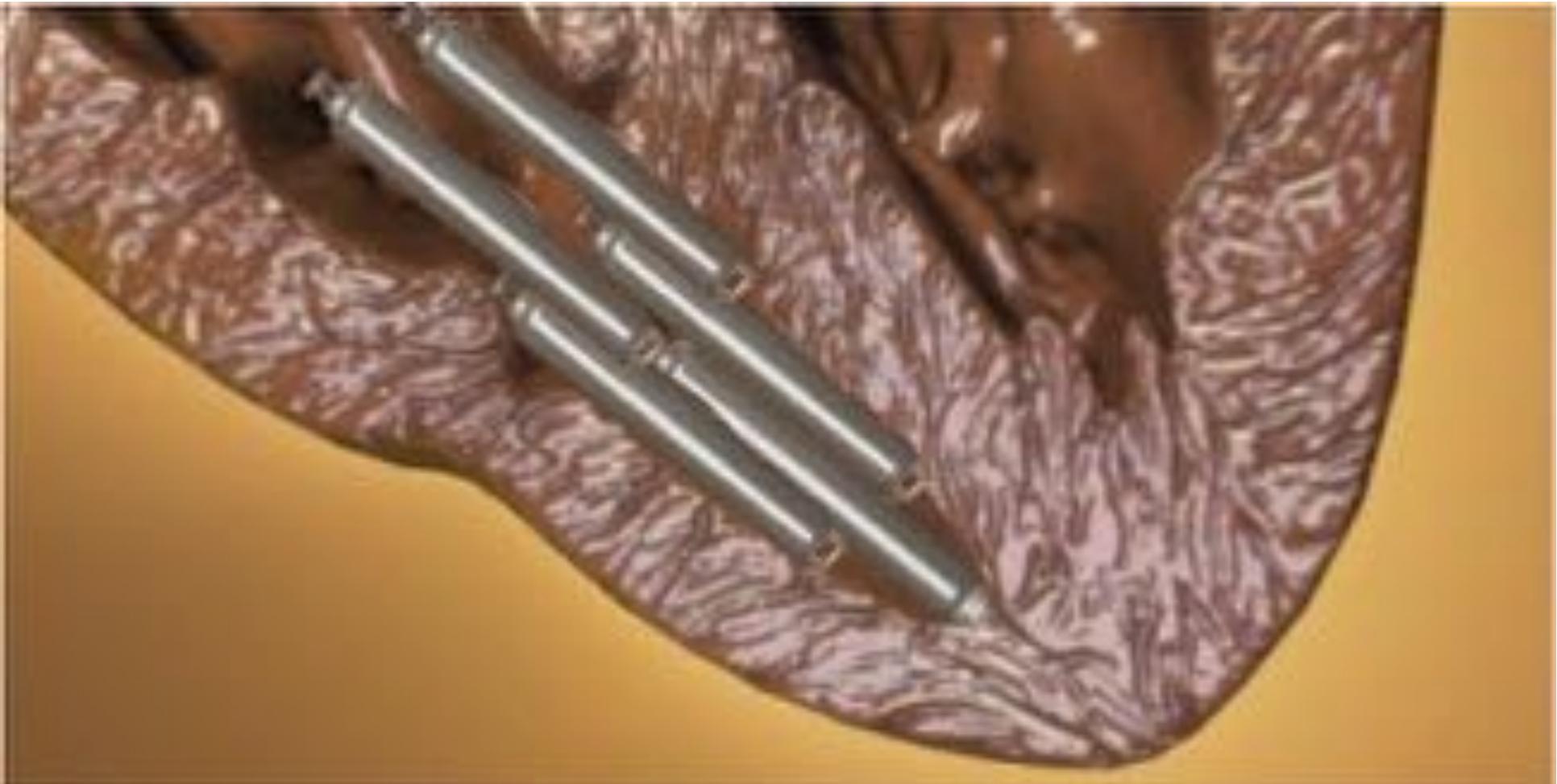
Les limites: que faire en fin de vie du dispositif ???



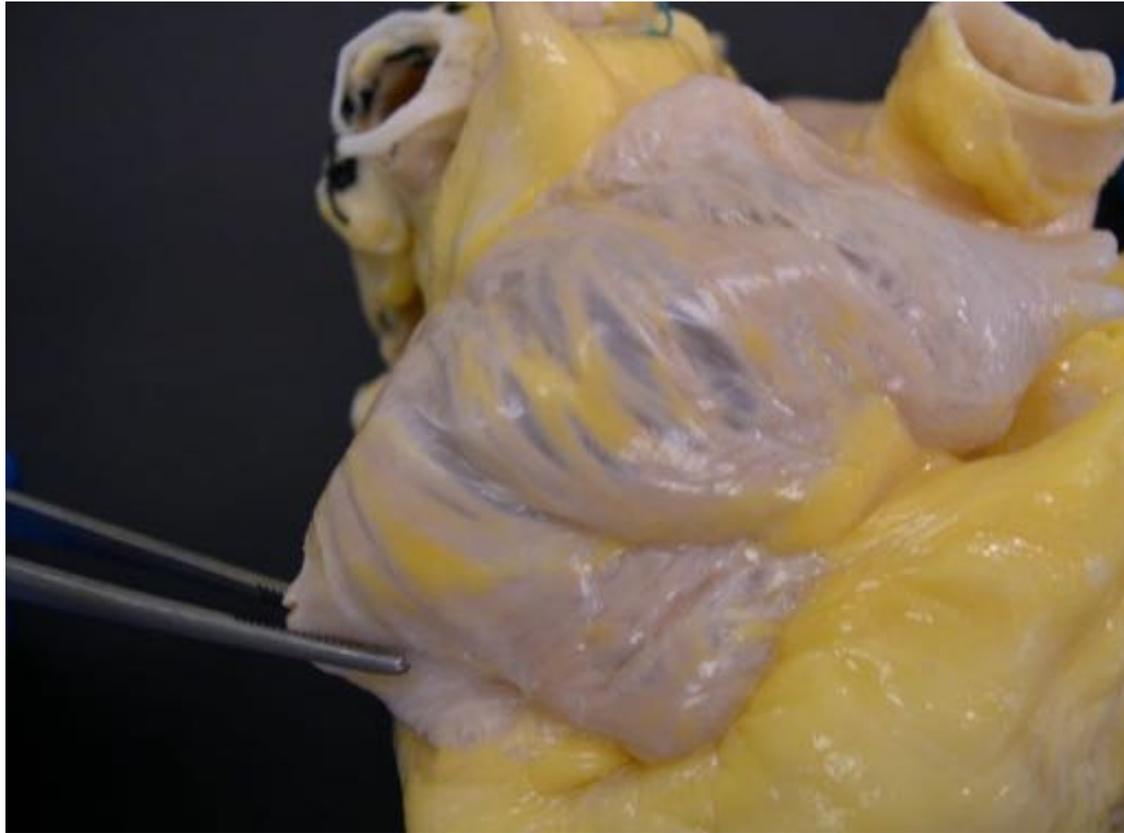
Les limites: que faire en fin de vie du dispositif ???



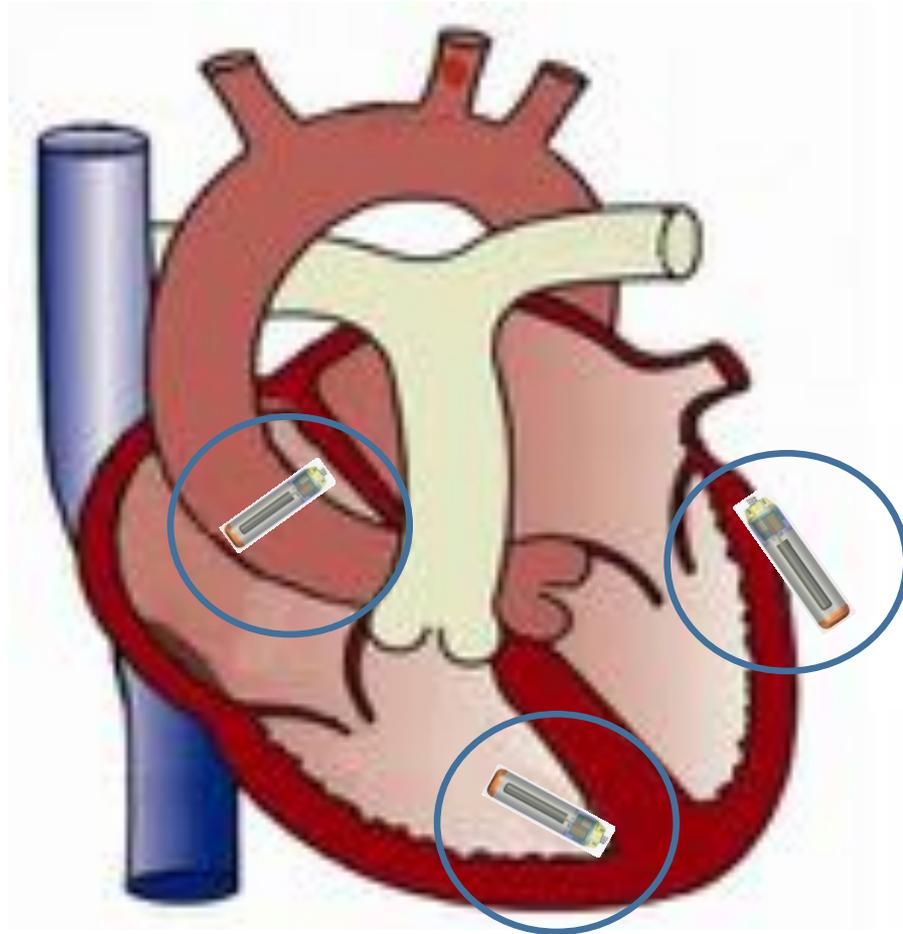
Les limites: que faire en fin de vie du dispositif ???



Les limites: sonde auriculaire? Sonde VG?



Perspectives ...



Perspectives ...

United States Patent [19]

Bilitch

[11] 4,256,115

[45] Mar. 17, 1981

[54] LEADLESS CARDIAC PACER

[72] Inventor: Michael Bilitch, Los Angeles, Calif.

[73] Assignee: American Technolgy, Inc., Northridge, Calif.

[21] Appl. No.: 111,576

[22] Filed: Jan. 14, 1980

Related U.S. Application Data

[30] Continuation-in-part of Ser. No. 112,566, Dec. 28, 1978, abandoned, and a continuation of Ser. No. 931,963, Oct. 2, 1978, abandoned.

[51] Int. Cl.² A61N 1/04

[52] U.S. Cl. 128/419 P; 178/701

[54] Field of Search 128/419 B, 419 F, 419 FG, 128/419 PS, 784, 792

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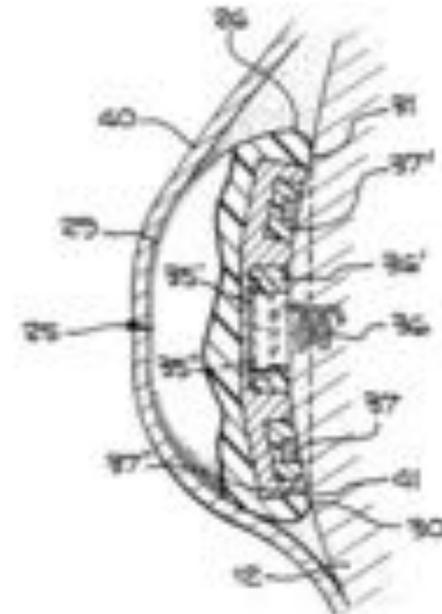
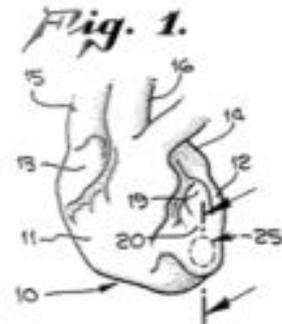
Scheiber et al. "Transactions of the American Society for Artificial Internal Organs", vol. 10, 1964, pp. 306-309.

Primary Examiner—William E. Kamm
 Attorney, Agent or Firm—Vernon D. Bricker

[57] ABSTRACT

A leadless battery operated cardiac pacemaker is embodied in a small disc-like case for attachment beneath the pericardium directly to the heart muscle. One electric lead is coiled and serves also as a means for both physically and electrically attaching the case to the heart muscle. The other lead surrounds the one lead and is drawn into surface to surface electric contact with body tissue to close the circuit. Solid state redundant circuitry makes provision to serve as a safety feature against possible malfunction.

9 Claims, 4 Drawing Figures



Restoring the rhythm of life

Comment stimuler une Cardiopathie congénitale...

Adverse impact of chronic subpulmonary left ventricular pacing on systemic right ventricular function in patients with congenitally corrected transposition of the great arteries



Wee Tiong Yeo ^{a,b,1}, Julian W.E. Jarman ^{a,1}, Wei Li ^{a,1}, Michael A. Gatzoulis ^{a,1}, Tom Wong ^{a*,1}

A B S T R A C T

Background: Patients with congenitally corrected transposition of the great arteries (ccTGA) are at high risk of heart block requiring subpulmonary left ventricular (LV) pacing. Long-term right ventricular (RV) pacing in congenitally normal hearts is associated with LV dysfunction. We examined the effects of univentricular subpulmonary LV pacing on the systemic RV in a ccTGA cohort.

Methods: ccTGA patients with two echocardiographic studies at least 6 months apart were included. Records of 52 patients, 22 with pacing, were retrospectively reviewed. Seven patients with biventricular pacing were included for comparison.

Results: The LV-Paced Group experienced deterioration in the RV fractional area change (RVFAC) (28.7 ± 10.0 vs. $21.9 \pm 9.1\%$; $P = 0.003$), systemic atrioventricular valve regurgitation ($P = 0.019$) and RV dilatation (end-diastolic area 32.7 ± 8.7 vs. 37.2 ± 9.0 cm²; $P = 0.004$). There was a corresponding deterioration in NYHA class ($P = 0.013$). Multivariate Cox regression analysis showed that pacing was an independent predictor of deteriorating RV function and RV dilation (hazard ratio 2.7(1.0–7.0) and 4.7(1.1–20.6) respectively). None of these parameters changed significantly in the Un-paced Group. The CRT Group showed improvement in RVFAC (22.0% to 30.7% ($P = 0.030$) and NYHA class ($P = 0.030$), despite having lower baseline RVFAC (22.0 ± 5.7 vs. $31 \pm 9.7\%$; $P = 0.025$) and greater dyssynchrony (RV total isovolumic time 13.4 ± 2.1 vs. 9.3 ± 4.2 s/min; $P = 0.016$) when compared to the Un-Paced Group.

Conclusions: Univentricular subpulmonary LV pacing in patients with ccTGA predicted deterioration in RV function and RV dilatation over time associated with deteriorating NYHA class. Alternative primary pacing strategies such as biventricular pacing may need consideration in this vulnerable group already highly prone to mortality from systemic RV failure.

Effects of Nonsystemic Ventricular Pacing in Patients with Transposition of the Great Arteries and Atrial Redirection

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TABLE 2
 Echocardiographic Results

	Nonpaced (n = 35)	Paced (n = 11)	P-Value
RV ejection fraction (%)	44 ± 10	39 ± 7	0.04
RV shortening fraction (%)	33 ± 10	27 ± 11	0.04
RV Tei-index	0.48 ± 0.2	0.58 ± 0.2	ns
RV dP/dt _{max} (mmHg/s)	1,024 ± 318	891 ± 470	0.04
S' of the tricuspid annulus (cm/s)	8 ± 2	7 ± 2	ns
Tricuspid regurgitation (mm ²)	8 ± 9	14 ± 14	ns
Interventricular delay ((ΔT _r RV-LV); ms)	31 ± 17	99 ± 100	<0.001
Right intraventricular delay ((ΔT _r RV-IVS); ms)	23 ± 11	70 ± 29	<0.001

TABLE 3
 Results of Exercise Stress Testing

	Nonpaced (n = 35)	Paced (n = 11)	P-value
Peak VO ₂ (mL O ₂ /kg/min)	26 ± 7	22 ± 6	0.04
VE/VCO ₂ slope	32 ± 7	35 ± 5	Ns
Oxygen pulse (mL O ₂ /beat)	10.9 ± 3.2	8.9 ± 1.5	0.05
Maximal exercise (W)	120 ± 32	100 ± 30	0.05
Circulatory power (mmHg mL O ₂ /min/kg ²)	4,540 ± 1,665	3,213 ± 1,094	0.02

Values are presented as means ± SD.

Conclusions: Long-term pacing of the nonsystemic ventricle in patients with atrial switch for TGA was associated with significantly impaired functional status, exercise capacity, and systemic ventricular function. (*J Cardiovasc Electrophysiol*, Vol. pp. 1-5)

Impact of pacing on systemic ventricular function in L-transposition of the great arteries

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Methods: We performed a retrospective review of all patients with a diagnosis of ccTGA who underwent pacemaker insertion. From 1993 to 2014, 53 patients were identified from the cardiology database and surgical records.

Central Message

All patients with congenitally corrected transposition of the great arteries who develop heart block should undergo primary biventricular pacing, as this therapy appears to prevent development of late-onset systemic ventricular dysfunction.

Perspective

Congenitally corrected transposition of the great arteries (ccTGA) is associated with a high incidence of atrioventricular nodal block due to unique conduction pathway characteristics, associated defects, and surgical interventions. The impact of univentricular versus biventricular pacing (BiVP) on systemic ventricular function remains poorly understood. This study demonstrates univentricular pacing is associated with late-onset systemic ventricular dysfunction, whereas primary BiVP preserves systemic ventricular function in patients with ccTGA. Primary BiVP should be considered in all patients with ccTGA who develop heart block.

(J Thorac Cardiovasc Surg 2016;151:131-9)

Indication stimulation BiV ou syst V sur VD systémique



Chez les patients appareillés et nécessitant une stimulation permanente

améliorer le statut fonctionnel
et éviter une altération progressive de la fonction du VD systémique



Primo implantation

en vue de préserver leur statut fonctionnel et hémodynamique

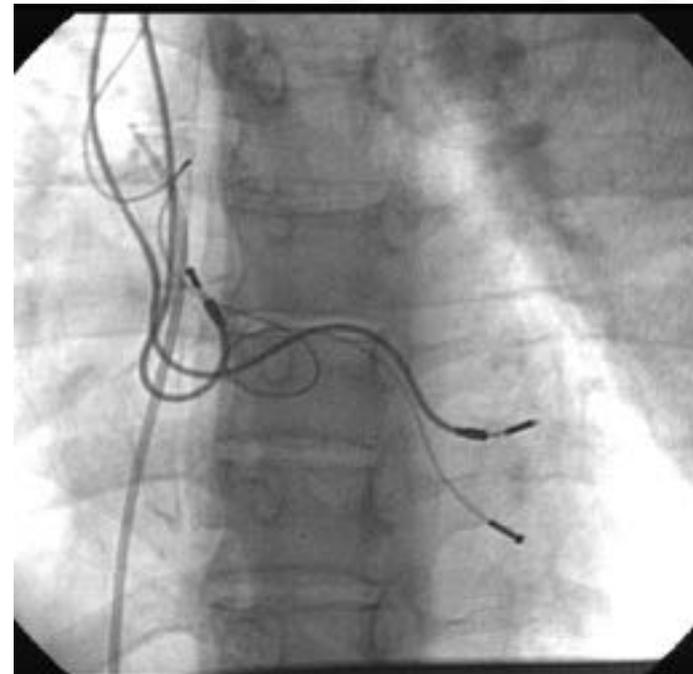


Pas de données pour proposer la stimulation BiV comme traitement de la dysfonction du VD systémique symptomatique ou débutante

Nécessite études multicentriques

Comment implanter

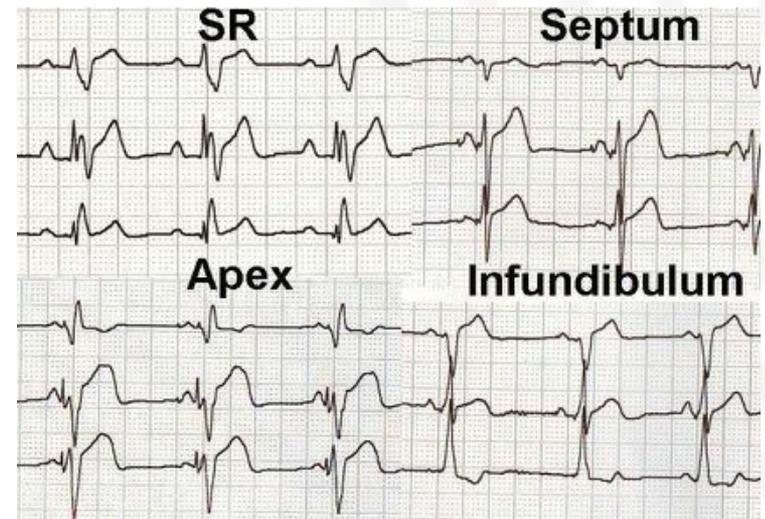
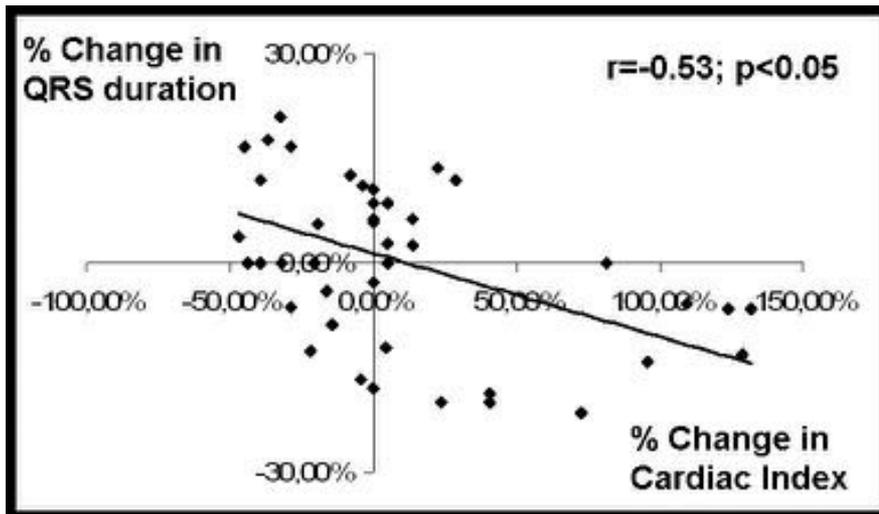
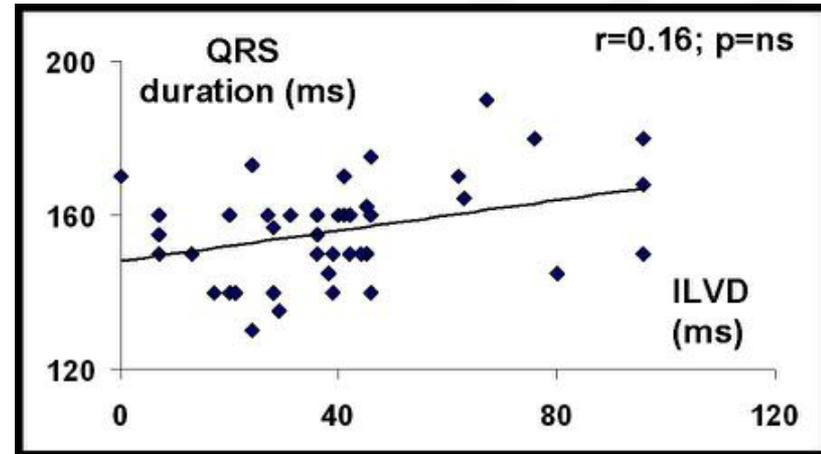
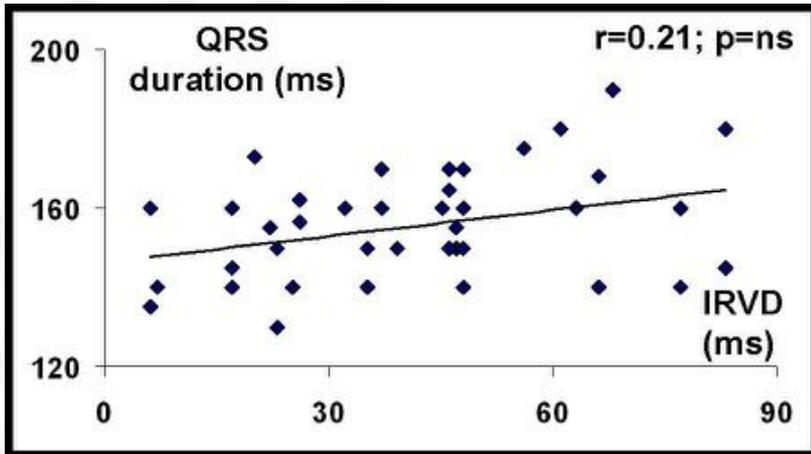
- Difficile ++
 - Stimuler le V systémique
 - Plusieurs approches :
 - Endocavitaire : mais sinus coronaire (souvent impossible accès)
 - Epicardique : chirurgie
 - Problème des sondes
 - Eviter sondes endocavitaires



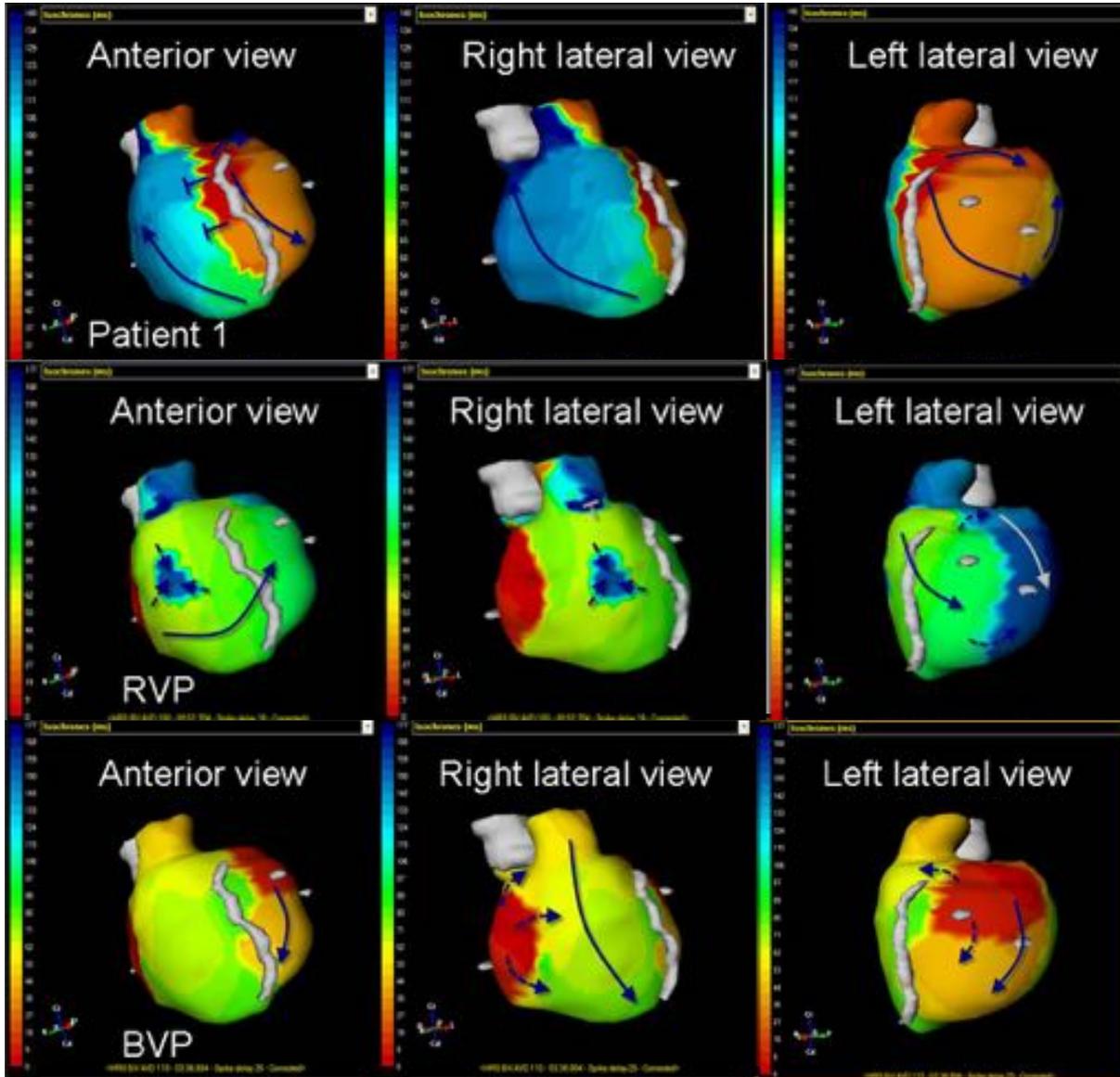
Tétralogie de Fallot : les questions

- BBDt +++ = asynchronisme
- La stimulation ne doit pas le majorer...
- Y a t'il un site ou un mode de stimulation qui puisse corriger cet asynchronisme ?
- Correction précoce de cet asynchronisme = remodelage (mécanique, électrique ?)

TOF RV pacing site



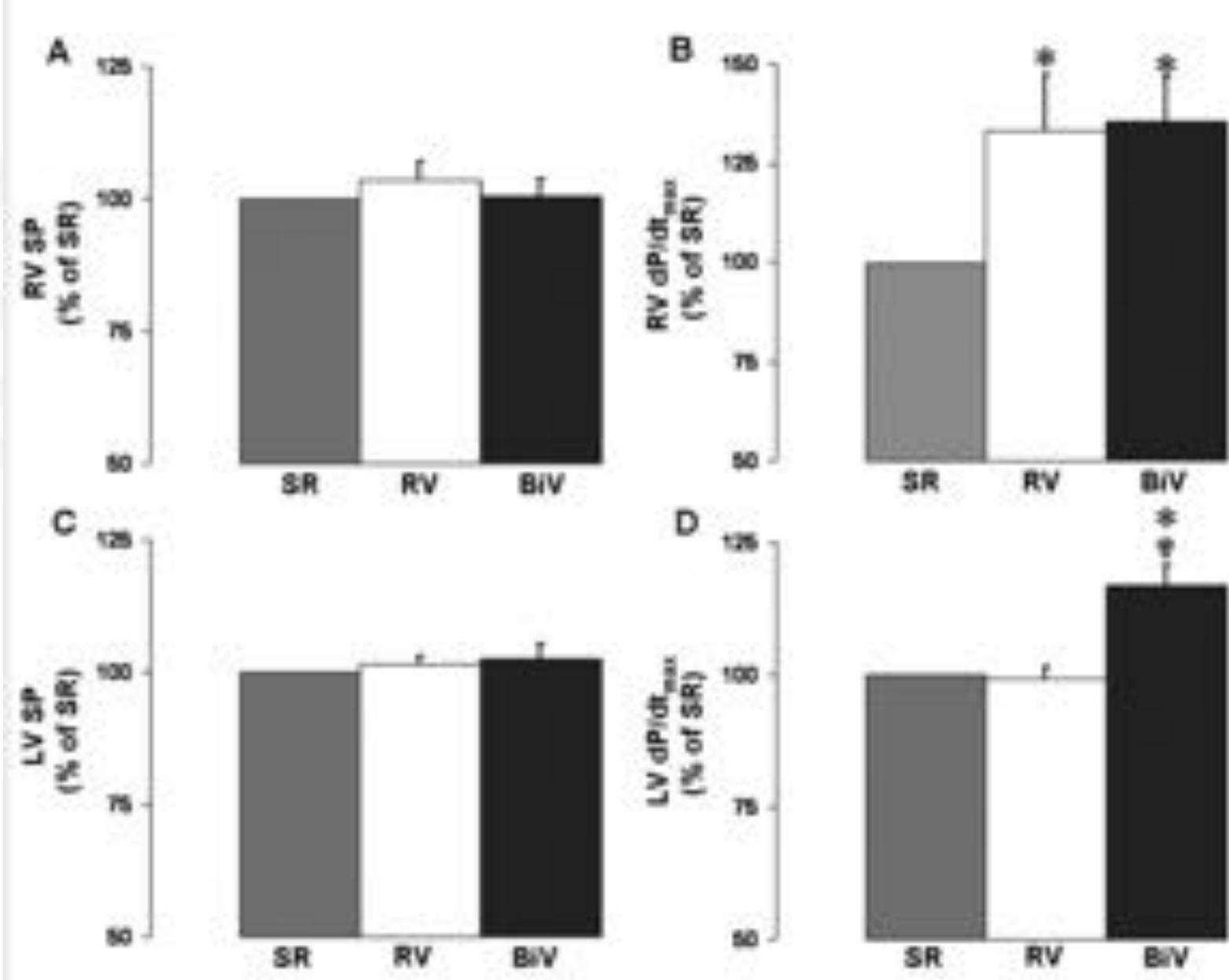
RS vs RV vs BIV Pacing in TOF / cardioinsight



 cardioInsight

Thambo JB, Int J Cardiol, 2011

TOF SR vs RV vs BiV



Thambo JB et al. Heart Rhythm. 2010

Conclusions

- Implanter en pensant à demain
- A ce jour implantation 100 % chir : enfant et adulte jeune...
- Besoin de développer des alternatives de stimulation **moins invasives, durables** qui **préservent** la fonction contractile